Prader-Willi Syndrome: Parent Perceptions of School, Professional, Social, and Informational Support, and Relations between Support, Child Behavior, and Stress

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Abstract

Prader-Willi Syndrome: Parent Perceptions of School, Professional, Social, and Informational Support, and Relations between Support, Child Behavior, and Stress
(Under the direction of Drs. Anne Wheeler and Rune Simeonsson)

Prader-Willi syndrome (PWS) is a rare genetic disorder that adversely impacts child development and health conditions, and is often associated with significant behavioral challenges. In particular, children with PWS typically exhibit extremely high levels of maladaptive behavior (e.g., excessive food seeking, hording, and binging; temper tantrums; aggression; stubbornness; and obsessive and compulsive behaviors). Child maladaptive behavior has been demonstrated as predictive of parent stress across many developmental disabilities and child conditions. However, prior to this study, there have been few studies, specifically describing the child behavior and parent stress relationship for families with children with PWS. Research has also indicated that, for other developmental disabilities, parent perceptions regarding received support are correlated with reductions in parent stress. However, prior to this study, there has been no current research documenting parental perceptions of the type and frequency of support received in the PWS population, nor have there been any current studies exploring the complex relations between support, child maladaptive behavior, and parent stress in parents of children with PWS. This study addressed these noted gaps in the literature by: 1) investigating parent perceptions of the availability and helpfulness of educational, professional,

social, and informational support sources; 2) examining whether or not parents perceive some sources of support to be more helpful than others; and 3) investigating the relations between educational, professional, social, and informational support, challenging child behavior, and parent stress in a sample of 61 biological parents (predominantly mothers) of children with diagnosed PWS. Findings indicated that parent perceived several individual support sources as particularly helpful, such as spouses/partners and supports that were idiosyncratic to PWS (i.e., PWS Foundations, Associations, and Clinics). In addition, for support sources that were utilized by greater than 40% of the sample, parents did not perceive any support source category (i.e., educational, professional, social, informational) as more helpful than any other. Child maladaptive behavior was predictive of parent stress, and sources of support by category (i.e., educational, professional, social, and informational) were not predictive of parent stress.

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TABLE OF CONTENTS

LIST OF TABLES

Page

CHAPTER 1

Introduction

Prader-Willi syndrome (PWS) is a rare developmental disability that is caused by a loss of chromosomal material, or expression of that material, on the paternal leg of chromosome 15 (15q11-113). The disorder affects 350,000 to 400,000 individuals worldwide, and has an estimated prevalence ranging from 1 in 8,000 to 1 in 20,000 (Butler, Hanchett, & Thompson, 2006). Generally, the disorder is characterized by extreme behavioral challenges, as well as physical and health-related problems, global cognitive and developmental delays, and an increased risk for psychiatric difficulties, (Butler et al., 2006).

Behavioral and emotional problems are hallmarks of PWS. Research has indicated that individuals with PWS present with more impaired behavioral functioning than most other developmental disabilities (e.g., Down Syndrome, Autism, and mixed aetiology learning disabilities; Dykens & Kasari, 1997; Rosner, Hodapp, Fidler, Sagun, & Dykens, 2004). Behavioral characteristics of the syndrome typically include overeating, temper tantrums, aggression, food and non-food obsessions, sleep disturbances, skin picking, self injury, over-verbalization, hoarding, stealing, lying, and compulsive, repetitive, and ritualistic behaviors (Dykens & Kasari, 1997; Hiraiwai, Maegaki, Oka, & Ohno, 2007; Kundert, 2008; University of Michigan Health Systems, 2008; Whittington & Holland, 2004). Emotional difficulties are commonly present as well, including mood lability, anxiety, stubbornness, and rigidness (Butler et al., 2006; Dykens & Kasari, 1997; Kundert, 2008; Whittington & Holland, 2004).

Furthermore, difficulties with regulating emotions often intensify behavioral problems, and thus increase externalizing behaviors such as temper tantrums and aggression (Butler et al., 2006; Dykens & Kasari, 1997; Kundert, 2008; Whittington & Holland, 2004). Moreover, for some individuals with PWS, behavior and emotional symptoms meet criteria for psychiatric disorders (e.g., anxiety, mood disorders, obsessive-compulsive disorder, delusions and psychosis; Butler et al., 2006; Hiraiwai, et al., 2007). Given the substantial severity of negative behavioral and emotional symptomology common in PWS, parenting a child with PWS is likely to be perceived as placing high demands on caregivers (Hodapp, Dykens, & Masino, 1997; Wingren & Hansen, 2003).

Parent stress is described as a special, complex case of stress is experienced as a result of parenting responsibilities (Deater-Deckard, 2004). Research has indicated that levels of stress are higher for parents of children with developmental disabilities than for parents of children who have other chronic conditions or who are typically developing (Guralnick, Hammond, Neville, & Connor, 2008). In addition, research particularly has indicated that child disorders with serious behavioral challenges are correlated with higher levels of parenting stress (Deater-Deckard, 2004; Mitchel & Hauser-Cram, 2008; Neece & Baker, 2008; Sprat, Saylor, & Macias, 2007; Suarez & Baker, 1997; White & Hastings, 2004). Parents of children with PWS in particular have reported high levels of stress associated with their children's behaviors (Hodapp, Dykens, & Masino, 1997).

Because chronic stress has an adverse effect on physical and mental health and interferes with effective parenting it has been suggested that methods for reducing parenting stress in the PWS population should be further examined by researchers (Deater-Deckard, 2004; Whittington & Holland, 2004). Support of all types has been identified as predicting reduced stress,

facilitating better coping, and leading to better child and parent outcomes (Seligman & Darling, 2007). More specifically, research on families with children with developmental disabilities and maladaptive behavior has indicated that social, educational, and professional supports are related to reduced parent stress (Deater-Deckard, 2004; Dunst, Jenkins, & Trivette, 1988; Green, Furrer, McAllister, 2007; Guralnick, Hammond, Neville, Connor, 2008; Hassall, Rose, & McDonald, 2005; Plant & Sanders, 2007; Seligman & Darling, 2007; White & Hastings, 2004).

In light of the literature on the relationship between parent stress and support, researchers have suggested that parents of children with PWS be given intense and consistent support from social, educational, professional and informational support sources (Butler et al., 2006; James & Brown, 1993; Wigren & Hansen, 2003; Whitman; 2006; Wyatt, 2006; van den Borne, van Hooren, van Gestel, Rienmeijier, Fryns, J. P., et al, 1999). Prior to this study, the relationship between perceived support and parent stress for parents of children with PWS has been minimally explored. Moreover, there has been a lack of current research that explores the types of support these families report receiving and the perceived effectiveness of those supports.

Hodapp et al. (1997) explored parent perceptions of social and professional support for families of child with PWS in a limited manner (i.e., with one open ended-free response questions). Results from that study suggested that when asked to list important supports, parents of children with PWS primarily listed "social" support sources, primarily friends and family members, as important. In addition, professional support sources were not listed by the majority of participants. Using survey methods that elicited responses regarding 67 individual support sources across four categories of support, the current study explored in greater depth the availability and perceived helpfulness of specific support sources. In addition, the associations between support, child maladaptive behavior, and parent stress were examined.

CHAPTER 2

Review of Literature

This chapter provides a brief overview of Prader-Willi Syndrome, describes the behavioral phenotype associated with PWS, and reviews the current literature on parenting stress in the context of child maladaptive behavior. Current literature on support for families with children with disabilities is also summarized. In particular, educational, professional, social, and informational (written and electronic) sources of support are discussed with regard to potentially decreasing parenting stress for parents of children with PWS. Finally, a discussion of previous studies exploring parental satisfaction with support and potential barriers to support is included. This chapter provides important background information relative to this study, however additional information, including a detailed review of the etiology of PWS and medical and developmental outcomes can be found in Appendix A.

Prader Willi Syndrome

Prader-Willi syndrome (PWS) is a rare and complex developmental disorder that affects 350,000 to 400,000 individuals worldwide, and has an estimated prevalence between 1 in 8,000 and 1 in 20,000 (Butler, Hanchett, & Thompson, 2006). PWS is caused by a loss of chromosomal material on the paternal leg of chromosome 15, in bands 11 through 13 (Whittington & Holland, 2004). This missing information has been found to result from three distinct genetic causes, leading to three subtypes with slightly different phenotypic presentations. The three subtypes, based on malfunctions within pre-expression genetic processes are: 1) deletions and

translocation, caused by a random mutation on the paternal leg of chromosome 15; 2) maternal uniparental disomy (UPD), where the child receives two copies of the maternal chromosome 15; and 3) imprinting defects (Buiting & Horsthemke, 2006), where the paternal chromosome carries a maternal imprint. Although the three genetic subtypes result in slightly different phenotypes, all subtypes have been described as resulting in significant behavioral challenges (Butler, Hanchett, & Thompson, 2006).

PWS occurs across all ethnicities and races, but is reported as occurring most often in Caucasians (Butler et al., 2006; Kundert, 2008). Just a decade ago, diagnosis did not occur until more severe symptoms, such as obesity, were observed (Butler et al., 2006; McCandless & Cassidy, 2006); however, the current diagnostic statistics show a trend towards earlier diagnosis (Kundert, 2008; Wigren and Hansen, 2003). Current data indicate that 29% of diagnoses are made by the age of one, 53% are made by the age five, and 89% are diagnosed by the age of seven (Kundert, 2008; Wigren and Hansen, 2003).

Physical and Development Characteristics

The physical characteristics of PWS include obesity, growth retardation, fair skin, small hands and feet, short stature, and dysmorphic facial features (i.e., narrow face, almond shaped eyes, and small mouth with thin upper lip and down-turned corners; Kundert, 2008; State & Dykens, 2000; University of Michigan Health Systems, 2008). Other features include hypotonia (weak muscles and low muscle tone), fatigue, and strabismus (i.e., cross eyes; Kundert, 2008; State & Dykens, 2000; University of Michigan Health Systems, 2008). Furthermore, hypoganadalism (i.e., low levels of sex hormones) and delayed or incomplete gonadal maturation are almost universally present (Crinò et al., 2003; Kundert, 2008).

Medical Challenges

Obesity is a primary health concern for adolescents and adults with PWS due to the presence of hyperphagia, (i.e., an insatiable appetite). PWS is noted as the most common form of obesity rooted in a genetic cause (Butler et al., 2006), and appetite and food problems are so pervasive that researchers have described PWS as a physiologically driven "eating disorder" (Greenswag & Alexander, 2006). Weight problems are exacerbated by short stature, low metabolic rate, impaired emesis (i.e., ability to vomit), and decreased need for calories (i.e., needing 40% to 70% fewer calories than typical peers; Butler et al., 2006; University of Michigan Health Systems, 2008). Obesity is more likely to be life-threatening if trained caregivers are not in place to manage overeating behaviors (Butler et al., 2006).

Serious medical problems result from obesity for these individuals. Complications such as high blood pressure, diabetes, cellulitis, hypoventilation, and chronic venus insufficiency (leading to ulcers and sores on legs and feet) often develop (University of Michigan Health Systems, 2008). Moreover, heart failure, hypertension, thrombophlebitis (vein inflammation related to blood clots), chronic leg edema, orthopedic difficulties, and Type 2 diabetes mellitus occur frequently (Butler et al., 20006; University of Michigan Health Systems, 2008). In addition, skin ulcers, sleep apnea impaired respiratory function, and various endocrine disturbances are a few of the other obesity-related problems that can manifest (Butler et al., 20006; University of Michigan Health Systems). However if obesity and overeating are controlled (i.e., forced exercise and extreme measures to monitor weight, including locking up all unauthorized food and food like-items), few serious health issue manifest, and life expectancy

can be similar to those with mild intellectual disabilities (Butler et al., 2006). Managing PWS overeating behaviors involves the implementation of intensive behavior management strategies by caregivers. Professional support sources such as pediatricians, medical specialists, dieticians, psychologists, PWS Foundations, and parent support groups are often recommended for aid in managing food-related behaviors, obesity, and medical complications for families with children with PWS (Butler, Hanchett, & Thompson, 2006; Whitman, 2006).

Cognitive and Academic Functioning

PWS is also characterized by intellectual and academic impairment (Butler et al., 2006). Generally children and adolescents with PWS exhibit lower intellectual functioning than their same-aged, typically-developing peers, with studies consistently suggesting average IQ scores in the mildly impaired range (55-70), or 40 points below the typically developing population (Butler et al., 2006; Whittington & Holland, 2004; Whittington, Holland, Webb, Butler, Clark, et al., 2006). However, these statistics fail to demonstrate the heterogeneous nature of cognitive functioning for this population (Butler et al., 2006). Generally speaking, approximately one third present with standard IQ scores within the 70 to 100 range, approximately two-thirds present with scores in the 50 to 70 range; and roughly 5% are indicated as functioning in the severe and profound range (Butler et al., 2006; Kundert, 2008; Whittington et al., 2004; Whitman & Thompson, 2006). Although children with PWS have cognitive challenges, research has demonstrated that parenting stress in parents of children with PWS is less related to the child's developmental or cognitive delays as it is to behavioral challenges (Hodapp, Dykens, & Masino, 1997).

Academic functioning is also impaired for those with PWS. Studies have found discrepancies between IQ and achievement scores that reflect significantly lower academic scores

(i.e., over one standard deviation) compared to indicated cognitive abilities (i.e. abilities indicated by IQ scores; Whittington et al., 2004). Downward discrepancies from IQ to achievement scores, referred to as "underachievement," have been identified for both general achievement variables as well as for specific academic areas (i.e., arithmetic, tasks involving social cognition or auditory processing, and academic skills that use short-term memory; Kundert, 2008; Whittington et al., 2004). Along with organic learning disabilities, interference by food- and non-food related behavioral problems have been implicated as facilitating underachievement (Kundert, 2008). A detailed review of cognitive and academic functioning in PWS can be found in Appendix B.

Phase I and II

The syndrome has two distinct phases that manifest, with the extreme behavioral problems manifesting in Phase II during early childhood. In Phase I infants and toddlers with PWS display extreme hypotonia, and often have weak suck reflex, leading to poor feeding, failure to thrive conditions, and frequent feeding tube interventions (Butler et al., 2006). Usually, Phase I begins to ebb around the end of the first year, and is typically followed by a short period of resolution of feeding difficulties and slow upward development (i.e., two to three years; McCandless & Cassidy, 2006). Next, behavioral problems begin in Phase II, with the onset of excessive appetite combined with a lack of hunger satiation (McCandless & Cassidy, 2006). Phase II symptoms include obsession with food and non-food objects, foraging and hording of food, intense temper tantrums and episodes of loss of emotional control, repetitive questioning, difficulty with transitions, and excessive need for routine and structure (McCandless & Cassidy, 2006). Unless intense food-related interventions are utilized (i.e., heavily monitored diet and

locking up off-diet food), obesity frequently develops, further compounding medical, health, and developmental challenges (Butler et al, 2006; Kundert, 2008).

Behavioral, Emotional, and Psychiatric Functioning

Studies have found that PWS stands out among developmental disabilities regarding the severity of maladaptive behavioral phenotypes (Dykens & Kasari, 1997; Oliver, Woodcock, & Humphreys, 2009; Rosner, Hodapp, Fidler, Sagun, & Dykens, 2004). Infants with PWS are generally noted as being happy, affectionate, compliant, and friendly (Kundert, 2008); however in early childhood (i.e., ages 2 to 5 years), significant behavioral problems emerge, with severely maladaptive behaviors becoming a key feature of the disorder (Kundert, 2008). More specifically, when compared to those with Down Syndrome (DS), Williams Syndrome (WS), and Nonspecific Intellectual Disability (NID), those with PWS demonstrate greater social impairment and more severe externalizing and internalizing behavior and mood problems (Dykens & Kasari, 1997; Rosner, Hodapp, Fidler, Sagun, & Dykens, 2004). Those with PWS have been found, cross-culturally, to demonstrate atypically high levels of overeating, food and non-food obsessions, sleep disturbances, skin picking, self injury, over-verbalization, hoarding, stealing, lying, and compulsive, repetitive, and ritualistic behaviors (Dykens & Kasari, 1997; Hiraiwai, Maegaki, Oka, Ohno, 2007; Kundert, 2008; University of Michigan Health Systems, 2008; Whittington & Holland, 2004). In addition, Rosner et al., (2004) found that although social competence abilities increased with age for the DS and WS groups, no age-related improvement was found for those with PWS. Moreover, significant social impairment and deficits in social

cognition are often present for those with PWS (Milner, Craig, Thompson, Veltman, Thomas, et al., 2005).

Maladaptive behaviors are often exacerbated by the emotional challenges and impulsivity, which also emerge as salient characteristics. More specifically, the characteristics of emotional dysregulation, negative emotionality, stubbornness, and rigidness (e.g., unwillingness to try new experiences and difficulty dealing with change in routine) often give rise to temper tantrums, aggression, noncompliance, defiance, and argumentativeness in the presence of routine school and familial expectations (Butler et al., 2006; Dykens & Kasari, 1997; Kundert, 2008; Whittington & Holland, 2004). In some cases, the severity of emotional and behavioral challenges reaches diagnostic levels, and psychiatric symptoms such as anxiety and mood disorders, (e.g., obsessive-compulsive disorder and depression), delusions, and psychosis can occur (Butler et al., 2006; Hiraiwai, et al., 2007). Furthermore, obesity combined with compulsive and sluggish behaviors can lead to being targeted for bullying by peers, which can, in turn, aggravate emotional and psychiatric difficulties (Dykens & Kasari, 1997). Behavioral, social, and emotional difficulties seem to worsen with age, as adolescents and adults with PWS are more prone to severe depression, anxiety, and psychosis than children with PWS (Kundert, 2008).

Additionally, emotional and behavioral problems become more intense in situations involving food for those with PWS. Food restricting practices are basic to proper physical care of those with PWS, and locking up unauthorized food and heavy monitoring of food access is typical (Whitman, 2006). However, in the presence of even mild food restrictions extreme behaviors such as food stealing, foraging, and hording, excessive secret binging, and even engaging in pica and eating unpalatable food (e.g., frozen food and food in garbage) can occur

(Butler et al., 2006; Young, Zarcone, Holsen, Anderson, Hall, et al., 2006). In addition behaviors towards caregivers surrounding the topic of food is often characterized by argumentativeness, temper tantrums, manipulation, and lying (Dykens & Kasari, 1997). Overall, a very large portion of time is often spent in food-seeking behaviors by those with PWS. Typically, intense behavioral interventions are recommended for the management of these problems; however, behavioral challenges are often resistant to intervention (Kundert, 2008).

Pharmacological interventions have often been utilized as part of a treatment regimen with minimum effectiveness (Whitman & Jackson, 2006). Anti-depressant, anti-psychotic, and mood stabilizing medications have frequently been prescribed, but weight gain side-effects have sometimes precluded the use of these drugs. (Kundert, 2008). Although psychotropic medications have demonstrated some effect on abnormal behaviors, no medication has been found to effectively treat hyperphagia (Butler et al., 2006; Whitman & Jackson, 2006).

Behavioral problems not only hinder adaptive functioning in those with PWS, but also have a profound impact on caregivers (Hodapp et al., 1997). For example, because children with PWS typically display high levels of child manipulation and low social skills, the parent-child relationship can be perceived as less emotionally enjoyable and more stressful than what is found in non-PWS parent-child relationships (Hodapp et al., 1997). Due to such severe behavioral challenges, PWS researchers acknowledge that parents or caregivers likely require support to implement necessary food management and behavioral interventions (Butler et al., 2006, Dykens & Kasari, 1997; Kundert, 2008). In addition, researchers and clinicians recommend that parents of young children find respite services, and that parents of older children find access to inclusive supervised activities during weekends and school breaks (Butler et al., 2006; Kundert, 2008). Such recommendations reinforce the notion that the disorder can facilitate atypically high levels

of stress in parents of children with PWS. In summary, PWS poses a plethora of unique behavioral challenges for those who provide support for children with the disorder and their families.

Parent Stress, Coping, and Support

Stress

Stress is a common human experience that involves significant psychological and physiological reactions which occur in response to a real or perceived aversive event or stimulus (Deater-Deckard, 2004). The autonomic nervous system (ANS), which aids in controlling several involuntary biological functions (e.g., heart rate, digestion, respiration rate, salivation, perspiration, diameter of the pupils, micturition), plays a large role in the physiology of the stress reaction (Deater-Deckard, 2004). The ANS is designed to maintain homeostasis in the bodily systems which it regulates. However, perceptions of events as stressful by the mind and emotions commence a temporary modification of the ANS functioning and a series of reactions in the body's sensory neural pathways, limbic system, hypothalamus, and neurotransmitters (Deater-Deckard, 2004). The body also releases stress related hormones (e.g., andrenocorticotrophic (ACTH), cortisol, thyroxin, oxytocin, vasopressin, epinephrine, and norepephrine) into the blood stream (Deater-Deckard, 2004). All these physiological changes prepare the body and mind for coping with the stressful event through increased tension and strength in the muscles, constriction of blood vessels, increased heart rate and breathing, ceasing of digestion, and several other physiological changes (Deater-Deckard, 2004). This physiological process moves the body out of homeostasis; however, ANS is only equipped to maintain functioning outside of homeostasis occasionally and only for short periods of time. The ANS is not equipped to maintain frequent or long term stressful physiological states. In other words, the human body is

not equipped to repeat this cycle continually, and thus chronic stress often results in the deterioration of physical and mental health due to over use of the stress response (Deater-Deckard, 2004).

Parent Stress and Raising Children with Disabilities

Parent stress is described as a special, complex case of stress. Deater-Deckard (2004) states that parenting stress differs from other types of stress in that in parents experience total responsibility for completely dependent others' survival and well-being. The author notes that although the demands of parenting are varied, they can be summarized as adapting to a child's individual attributes and needs, as well as fulfilling perceived social parenting roles. Furthermore, parent stress occurs when there is a discrepancy between parent perceptions regarding the demands of parenting a child and available resources for meeting those demands (Deater-Deckard, 2004). Hence, Deater-Decker (2004) proposes that parent perception of stressors can be just as important as actual stressful events, and perceptions of supports can facilitate lower levels of stress. She notes that, even for parents of typically developing children, parenting often requires continuous, immediate, and sometimes aversive tasks to be fulfilled (Deater-Deckard, 2004). She also notes that child needs, and thus parent demands, are often significantly intensified in children with disabilities (Seligman & Darling, 2007). Therefore, in parents of children with significant and severe special needs, such as children with PWS, stress-producing perceptions regarding the exorbitant demands of parenting tasks could be considered expected and realistic.

Currently well-accepted theories of parenting stress include the Parent-Child-Relationship Stress model (P-C-R), the Daily Hassles theory (DH), and the Ecological Model of Parenting Stress (Deater-Deckard, 2004, Seligman & Darling, 2007). Each of these theories can be viewed

as consistent with the ecological framework that Bronfenbrenner (1979) introduced, as they describe parenting stress in terms of the impact of interacting systems (e.g., the child, family, and community as systems). Although these theories have been conceptualized to describe parenting stress in the general population, the theories are thought to be useful in understanding parent stress in parents of children with disabilities (Deater-Deckard, 2004, Seligman & Darling, 2007).

The P-C-R model views stress as generated from aspects of parenting that arise from within 1) the parent (P); 2) the child's behavior (C); and 3) the parent-child relationship (R; Deater-Deckard, 2004). With the inclusion of the parent-child relationship element, stress is seen as bi-directional, as perceptions and behaviors by both parties interact. One notable feature of this theory is that it recognizes that elevations in perceived care giving stress can be found in families where children evidence atypically high needs due to developmental delays, such as children with PWS (Deater-Deckard, 2004). In addition, the theory accounts for relationally originated elements of stress, which can result from child emotional and behavioral characteristics that affect the child-to-parent relationship (Deater-Deckard, 2004).

Within the Daily Hassles (DH) theory, stress results from daily parenting "hassles," such as managing children's misbehavior (Deater-Deckard, 2004). In this theory, the severity and quantity of hassles accumulate to predict well-being and mental health outcomes for parents (Deater-Deckard, 2004). When viewed through the DH theory, the behaviors of children with PWS could be seen as presenting a high number and high severity of daily hassles due to the excessive challenges in functionality and constant need to manage behaviors (Whitman & Jackson, 2006).

Lastly Seligman & Darling (2007) use an ecological framework for viewing families of children with disabilities. Here families are embedded in the social structures of community

relationships and societal happenings and norms over time. Hence, parent stress occurs as a result of inadequate support at the family, community, and societal levels over time. The system framework helps explain the nested nature of relationships, and thus findings that parent stress that is facilitated by child disability adversely affects parent engagement with the child (Wheeler, Hatton, Reichart, & Bailey, 2007). The ecological framework indicates that stress could be reduced through interventions (e.g., support) that addresses any presenting child and family challenges, especially so for parents of children with severe impairments.

Taken together, the Parent-Child-Relationship Stress model (P-C-R) the Daily Hassles (DH) theory and the Ecological Model of Parenting Stress have implications for stress and parents of children with PWS. The P-C-R and DH models illustrate how aversive types of interactions (e.g., temper tantrums, moodiness, etc.) as well as the high quantity and level of severity of negative behaviors (e.g., food stealing, lying, repetitive talking, etc.) can produce parenting stress in parents of children with PWS. In addition, the P-C-R and Ecological models together indicate that parent perception of adequate support and resources from outside the family can help alleviate parent stress. Seligman & Darling (2007) suggest that families with sufficient resources, and abilities to use those resources, experience less stress.

Researchers have identified five areas that produce stress for parents of children with disabilities and chronic health conditions: 1) intellectual stress, or stress from lack of information regarding etiology, treatment, and prognosis; 2) instrumental stress, or lack of resources to incorporate the tasks necessary regarding a child's treatment and care into the family lifestyle; 3) emotional stress, resulting from anxiety and lack of resources to maintain adequate sleep, diet, and energy; 4) interpersonal stress, or distress in relationships with family members, friends, or

medical and educational personnel; and 5) existential stress, or difficulty with constructing meaning out of the experience of rearing a child with a disability (Seligman & Darling, 2007). This model suggests that accurate information about PWS specific to one's child and family, social support and practical help and professional support might be key elements in reducing parenting stress.

Research coincides with theoretical perspectives that parents of children with disabilities experience more stress on average than those with typically developing children. Despite some individual differences in parent coping styles, the positive relationship between child maladaptive behavior and parent stress has been established consistently across child age ranges, cultures, ethnicities, disorders, chronic physical conditions, developmental disabilities, levels of intellectual impairment, and social economic statuses (Green, Furrer, McAllister, 2007; Guralnick, Hammond, Neville, Connor, 2008; Hassall, Rose, & McDonald, 2005; Horton & Wallander, 2001; Mitchell & Hauser-Cram, 2008; Plant & Sanders, 2007; White & Hastings, 2004). For instance, higher levels of stress are indicated in general for parents of children with externalizing behavioral problems, with serious delays, and with learning disabilities (Deater-Deckard, 2004). Also, child behavior problems in the social skills domain are correlated with high maternal stress, over and above those with children with just behavior problems or impaired intellectual status (Neece & Baker, 2008). In a sample of parents of children with various special needs, parents experienced higher levels of stress from child behavior problems alone and child maladaptive behavior with intellectual disability, yet lower parent stress when children had only intellectual disability (Spratt, Saylor, & Marcias, 2007). Similarly, Beck, Hastings, and Daley (2004) found that for parents of children with intellectual and developmental disabilities, problematic behavior, and not poor adaptive functioning predicted parent stress. Research

indicates that for parents of children with intellectual disability, those with children with more severe impairment evidence higher stress levels (Deater-Deckard, 2004). In addition, studies generally indicate that stress increases over time for parents of children with developmental disabilities (Deater-Deckard, 2004; Seligman & Darling, 2007). Taken together, disability and parent stress are correlated, especially with regard to disabilities that include high levels of maladaptive behavior and severe over-all impairment.

Parent Stress and PWS

Although research indicates a positive correlation between child behavior problems and parent stress, little research has demonstrated this relationship specific to parents of children with PWS (Green, Furrer, McAllister, 2007; Guralnick, Hammond, Neville, Connor, 2008; Hassall, Rose, & McDonald, 2005; Horton & Wallander, 2001; Mitchell & Hauser-Cram, 2008; Plant & Sanders, 2007; White & Hastings, 2004), Research has indicated that in families of children with PWS, child behavioral problems correlate with parent stress, whereas obesity and intellectual disability do not (Hodapp, Dykens, & Masino, 1997). More specifically, Hodapp et al.'s study (1997) measured behavior with the Child Behavior Checklist (CBCL; Achenbach, 1991), and found that child behaviors captured by the "Other" category of the CBCL were most associated with parent stress (r = .60). Other domains of behavior were also correlated with parent stress: internalizing (r = .31); externalizing (r = .45); social (r = .38); and thought (r = .43), problems specifically. Furthermore, Whittington & Holland (2004) found that parents of children with PWS experience higher levels of stress and depressive symptoms, and seek more psychological treatment than parents of children with a mixed etiology of learning disabilities. Results from these studies suggest a need for more research examining the unique challenges faced by parents of children with PWS, as well as possible sources for stress reduction. The current study was

designed to address this need by expanding the literature describing challenging behaviors and parenting stress as well as delineating the sources of support found to be most helpful by parents.

Parent coping and support

Parenting stress is linked to child abuse, less effective parenting, less adaptive parent-child relationships, and poor parent health (Deater-Deckard, 2004; Spratt, Saylor, & Marcias, 2007; Wheeler, Hatton, Reichart, & Bailey, 2007); therefore, reducing parenting stress is of paramount importance for families. Reducing stress involves removing stressors externally (i.e. in the environment), or reducing the perception of the experience as negative (i.e., change the subjective experience of events), and therefore precluding activation of the physiologic stress reaction (Deater-Deckard, 2004). Coping with parenting stress can involve problem-solving-focused resources, which are useful in removing stressors that are under one's control, and emotion-focused resources, which can be used to change one's experience of stressors (Deater-Deckard, 2004).

Clinicians and medical providers have typically addressed the needs of families with children with disabilities through recommending help via various sources of support (Deater-Deckard, 2004; Grant & Whittell, 2000; Haveman et al., 1997; Ogletree, Fischer, & Shultz, 1999). In particular, care providers for those with PWS recommend support from the social, educational, professional (non-school affiliated), and informational sources for the child and family (Cassidy & Driscoll, 2009; James, & Brown, 1992; Kundert, 2008; Wyatt, 2006). Although the effectiveness of help and interventions from these sources of support are generally based on anecdotal information (Kundert, 2008), the model of care for families with children with PWS is typically presented as including multiple support services from all four noted sources of support (Butlter et al. 2006). Moreover, some research has indicated that, although the

need for support for families with children with disabilities is continuous, the perceived need for certain sources of support can change according to child age (Grant & Whittell, 2000; Haveman et al., 1997). Below, frequently recognized child and family support needs for families with children with PWS are discussed. The literature on the impact of support from educational, professional, social, and informational sources on parent stress and family functioning is also presented.

Support Needs of Children with PWS

Although the relationship of support and family outcomes has not been researched for families with children with PWS, many support needs have been identified across many domains of life, including health and medical, educational, and family domains.

Medical, health-related, and developmental support needs. The medical and healthcare support needs for children with PWS are typically significant. Medical interventions, especially those that address life-threatening early under-eating and latter hyperphagia and obesity, are often required. Moreover, as early as the second and third year of life, parents need guidance as they begin adjusting to the need for managing their children's weight and diet as hyperphagia sets in (Whitman, 2006, Whitman & Jackson, 2006). More specifically, parents need to acquire weight and food behavior management skills including implementing a strict diet and exercise plan; altering the physical environment to preclude access to restricted food; elimination of all extraneous avenues of obtaining food; and applying strategies for quelling child anxiety by keeping children informed regarding times and menus for upcoming snacks and meals (Whitman & Jackson, 2006; University of Michigan Health Systems, 2008). Other frequently needed medical interventions include growth hormone treatment, sleep studies and interventions, orthopedic interventions, and sex hormone replacement therapies (University of Michigan Health

Systems, 2008). Additionally, due to frequent speech and motor delays, speech, physical and occupational therapies are frequently recommended and sought (University of Michigan Health Systems, 2008).

Educational support needs. Children with PWS have complex and demanding needs within the school setting for which they need support (Whitman & Jackson, 2006; Wyatt, 2006). Although data on school services and PWS is sparse, Wyatt (2006) sited that poor understanding of PWS by school staff is a common parent complaint. He also noted reports that poor understanding of the disorder often lead to poor school services. Parents can often find themselves in the predicament of educating school staff while advocating for services for their children (Jackson & Brown, 1992; Wyatt, 2006). In addition, due to the popular "no-tolerance" policies regarding discipline in public schools, children with PWS also frequently need advocates within the school who understand the behavioral difficulties that PWS presents and can preclude harsh and inappropriate discipline for PWS-related behavioral infractions (Wyatt, 2006).

Family support needs. Expectations regarding family life need to be adjusted to the mental, physical, and behavioral abilities of the child with PWS. In particular, intense environmental modifications, such as rigid, specific, and consistent household rules and schedules, are often needed to prevent emotional and behavioral escalations (Whitman & Jackson, 2006). In addition, coping with child behavioral symptomology and manipulation is typically a constant challenge for parents (Butler et al., 2006; Kundert, 2008; Whitman & Jackson, 2006). Also, food restrictions for children with PWS can mean food restrictions for non-PWS siblings, which can introduce added challenges to balance within the family system. Moreover, unlike typically developing children who mature and eventually become less dependent on their parents, children with PWS will continue to be either partly or completely

dependent on their others for shelter, care, and medical, behavioral, and psychological management (Seligman & Darling, 2007). The direct financial cost of interventions, care, and support can also accumulate and impact family life. Given the manner in which PWS can impact family life on a daily- or long-term basis, families likely need education and support in developing methods for managing family life, (van Hooren, Widdershoven, van den Borne, & Curfs, 2002; Whitman, 2006, Whitman & Jackson, 2006).

In summary, PWS is condition that significantly challenges the individual and family system, and support that addresses child and family needs, especially with regard to improving child behavior and functioning, is likely needed. Next the literature regarding educational, professional, social, and informational support sources as means of meeting such needs are explored.

Sources of Support and PWS

In 1997, Hodapp et al. investigated the importance of certain support sources in regard to PWS families. They utilized an open ended survey question that addressed social and professional support over a short duration of time with 42 families with children with PWS. More specifically, he asked parents to "list the names of people who are important to you at this time in your life," and specified that parents considered social and professional connections with whom they had been in contact over the previous 4 to 6 weeks. Sources of support gathered via the responses to the open-ended question were generalized broadly into categories of "social" and "professional" supports. Using these methods, families in this study reported an average of 7.5 individuals who were sources of support, with 92% of the responses being categorized as "social" support, primarily by friends and family members. In addition, 67% did not list any professionals as "important" sources of support.

Educational support. Support sources within the educational setting for children with disabilities involves providing services that enable access a free and appropriate education in a least restrictive environment as described in the Individuals with Disabilities Education Act (IDEA; P.L. 108-446, 118 Stat. 2647). For children with disabilities, a free and appropriate education usually includes an Individualized Education Program (IEP), which specifies provisions such as modified curriculum, special school-based interventions, and school accommodations for the individual child. Chedd, Levine, & Wharton (2006) describe an IEP as the "blueprint of all aspects of that child's special education and the resulting services to be provided." Each child receiving services under IDEA must have an IEP team, which creates and supports the child's educational plan (P.L. 108-446, 118 Stat. 2647). IDEA 2004, Section 1414(d)(1)(B) stipulates that parents of children with disabilities are to be an integral part of this IEP team, sharing equal decision making power with the other team members. Appropriate educational support also involves other school personnel working effectively with parents and children, to facilitate an education that is appropriate for the child.

Due to the high levels of learning and behavioral problems that those with PWS demonstrate, the need for school support has been well-recognized by researchers and clinicians, and improved empirically based school support practices have also been a well recognized need (James & Brown, 1993; Kundert, 2008; Wyatt, 2006). Parents appear to be in agreement of the need for better support. For example, communications received by the Prader-Willi Association (USA) crisis intervention center indicate that, within the school setting, parents are seeking support regarding school advocacy and legal rights (Wyatt, 2006). However, very little research has been published on school support and children with PWS (Heinemann, 2008).The only known research on this topic was published in 1993 when James and Brown indicated that school

support was correlated with decreased parenting stress for parents of children with PWS; they noted that parent stress decreased with access to various services, including special education support (James & Brown, 1993). Recognizing the need for more current and comprehensive research in this domain, the PWS Association-USA recently initiated a call for research on "Schools and PWS" (Heinemann, 2008).

Despite the gap in research, PWS researchers recognize the need for school support for families of children with PWS in their clinical writings. For example, Whitman (2006) emphasized that families with children with PWS need intense support around educational issues, such as school-related services, case management, and interventions. Chedd et al., (2006) suggested that children with PWS have specific, unique, and evolving developmental and educational needs that need to be addressed in the school setting across developmental stages.

Regarding parents with children with developmental disabilities, Haveman, van Berkum, Reijnders, & Heller, (1997) reported that parents rated school support as the most essential type of support during the school age years. In another study, parents of children with autism in North Carolina indicated that school support services were the most germane and effective services utilized during the school years, and indicated that 90% of all support services they received came directly through their children's schools (Thomas, Morrissey, & McLaurin, 2007). Given that school support is correlated with reduced stress for parents of children with other developmental disabilities, and that parents of children with PWS are requesting better support in this area, exploring perceptions of the current nature and helpfulness of educational support sources for families with children with PWS is important.

Professional support. Professional support (i.e., community support of any type found outside of the schools) can include support from medical and mental healthcare providers,

professional associations and organizations, community care providers, and government-based agencies. Literature on developmental disabilities has indicated that professional support can be correlated with reduced parental stress. For example, research on parents of a general sample of children with developmental disabilities indicated that when age, IQ, and language development are controlled for, the relationship between child behavior and parent stress decreases with professional support (e.g., parenting information and practical aid regarding care giving; Guralnick, Hammond, Neville, Connor, 2008). Deater-Deckard (2004) adds that developmental knowledge and information communicated through professionals is necessary for empowering parents of children with developmental disabilities. She also notes that professional support is helpful in coping with stress from stigma or parent-perceived difficulties with meeting child needs (Deater-Deckard, 2004). Meta-analysis research indicates that effective professional support reduces parenting stress through employing strategies that improve parenting skills, support children's developmental functioning, and provide external support for parent-child relationships (Deater-Deckard, 2004). Mitchell and Hauser-Cram's research (2008) indicates that high levels of quality medical and health care services predict lower maternal stress and depressive symptoms for parents with children with developmental disabilities. Other researchers have indicated that parents of children with intellectual disabilities and psychopathology need support from the professional community that provides parent counseling, support in securing medical health care services, crisis support, material and practical aid, such as respite support, and information on available disability services and appropriate extracurricular activities (Deater-Deckard, 2004; Douma, Dekkar, & Koot, 2006).

Although current research is available with regard to professional support sources for families with developmental disabilities, only a few studies have addressed professional support

in regard to PWS. In 1993, researchers found that for parents of children with PWS, parent stress decreased with access to community and social services, especially respite care and special education support (James & Brown, 1993). Hodapp et al.'s (1997) study pointed to the lack of professional support for families by indicating that professionals only provided 8% of overall support for families regarding raising and caring for their children with PWS. In addition, the families in Hodapp's study reported an average of only three professionals involved in their children's healthcare, and 28 of the 42 families reported that no professionals were a part of the family support system.

More recently, Wigren and Hansen (2003) explored parent perceptions of support perceived as needed. More specifically, 42% of 58 parents with children with PWS reported desiring professional counseling for their children which addressed the impact of PWS on their children's experiences. Moreover, results indicated that parents were seeking information on accessing healthcare services. Van Hooren, Widdershoven, van den Borne, and Curfs (2002)'s research explored quality care regarding medical care and PWS, and reported that appropriate medical support services should include a strong physician-patient relationship that incorporates the child's wishes and facilitates a developmentally appropriate amount of self-determination and autonomy.

Despite the sparse research in this area that is specific to PWS, those who support families with children with PWS continue to advocate for professional support as a means to address the plethora of needs the syndrome presents. Recent authors have suggested that parents of children with PWS be provided with continuous intensive psychosocial and professional support to address the negative behavioral symptoms of PWS which often worsen with age (Wigren & Hansen, 2003; Wyatt, 2006). Whitman (2006) emphasizes the need for intense family support,

including professional support and advocacy in accessing and coordinating case management, medical, healthcare, and intervention services. Turnbull, Turnbull, Erwin and Soodak (2006) suggest that support should be available from community resource centers, family organizations, and other community professionals and should include within family support and parent training, parent-to-parent and family-to-family support. In addition, professional support for marital and family relationships, and parent training and support are also often noted as necessary (Whitman, 2006). Taken together, PWS researchers and organizations advocate for high levels of intense professional support for families with individuals with PWS, but little research is available on the subject that is specific to PWS.

Social support. Researchers have generally defined the construct of social support as emotional and practical support by individuals within one's social affiliations. The social network typically can be thought of as including family members, friends, neighbors, and acquaintances who can provide support, but do not act as professional care providers (Guralnick, 2008; Suarez & Baker, 1997; White & Hastings, 2004). Social support has been identified as helping with coping in general. For example, Olstad, Sexton, & Søgaard, (2001) found that the relationship between general life stressors and mental distress is buffered by social support, with the effect being even stronger for women.

Researchers have demonstrated a strong correlation between social support and decreased parent stress. Deater-Deckard, (2004) indicated that social support facilitates more effective coping for parents and enables parents to better meet the demands of stressful events . She proposed that social support decreases the experience of parent stress through facilitating feelings of competence and well-being, and thereby shoring up resilience factors which aid in successfully facing crisis. In particular, parents need supportive social relationships from which to glean

emotional support and receive empathy, and thus reduce aversive stress-driven emotions (Deater-Deckard, 2004). Meta-analysis studies indicate that relationship-focused-coping, which involves emotional support from social support sources, is necessary for reducing parenting stress, and for maximizing the effect of problem-solving-focused and emotion-focused coping resources (Deater-Deckard, 2004; White & Hastings, 2004). In addition, instrumental or practical social support, that is receiving practical assistance from family and one's social network, is also reported as necessary for reducing parent stress (Deater-Deckard, 2004).

Disability research indicates parents of children with more severely impairing conditions and with high levels of behavioral challenges need significantly greater amounts of support from social sources for effective coping with parent stress (Guralnick, Hammond, Neville, & Connor, 2008; Hodapp, Dykens, & Masino, 1997; Plant & Sanders, 2007; Seligman & Darling, 2007; Suarez, & Baker, 1997). As noted previously, in 1997, Hodapp et al. asked parents of children with PWS to support individuals who were "important" in their lives of the previous 4 to 6 weeks, and found that the names of parents' family members and friends were most often reported. For parents of children of all ages with various externalizing behavior challenges, perceived social support (i.e., perceived spousal support, marital adjustment, and global social support) has been shown to decrease parent stress (Guralnick, et al., 2008; Plant & Sanders, 2007; Suarez, & Baker, 1997). White & Hastings (2004) found that, for parents of children with intellectual disability, social support was more correlated with parent well-being than was support by professionals. Additionally, in mothers of preschoolers with developmental disabilities and behavioral challenges, social support has been correlated with decreased maternal stress (Plant & Sanders, 2007). Hassall et al. (2005) found that mothers of children with intellectual disabilities who perceived more social support (as measured by the Family Support

Scale; Dunst, Jenkins, & Trivette, 1988), experienced lower parenting stress (as measured by the Parent Stress Index: Abidin, 1995). Similarly, Boyd's meta-analysis (2002) indicated that parents of children with autism and who received social support were better able to emotionally relate to their children.

Social support has been well documented as correlated with decreased distress in parents of children with and without special challenges (Seligman & Darling, 2007). Moreover, social support from family members has been reported as the greatest need for those who parent children with developmental disabilities; however, community support is especially important for those without family support (Seligman & Darling, 2007). Conversely, the lack of social support has been identified as contributing to isolation, depression, and negative outcomes which are already frequently experienced by those with children with developmental disabilities (Deater-Deckard, 2004; Guralnick, 2008). In addition, research indicates that when further risk factors are present, such as low socio- economic status, social support is helpful in decreasing maternal anxiety and increasing positive parent-child interactions (Green, Furrer, McAllister, 2007).

Thus, research has indicated a correlation between social support and decreased stress for parents of children with disabilities, yet research has been inconclusive in regard to a correlation between support and decreased stress specifically for parents of children with PWS (James & Brown, 1993, Hodapp et al. 1997). Hence, the potential support and stress relationship is still in need of exploration in regard to parents of children with PWS. Furthermore, White and Hastings (2004) note that, although much research has addressed the impact of social support on parents stress for parents of children with disabilities, research on social support typically does not account well for availability of various sources of support. This study will address the question of

availability of four categories of sources support and provide current data on social support and PWS.

Printed and electronic informational support. Whereas the above discussed sources of support involve direct contact with human beings, printed and electronic informational support is information that can be read by the support recipient. This type of support can include books, pamphlets, journals, and internet sites. Research by van den Borne, van Hooren, van Gestel, Rienmeijier, Fryns, J. P., et al, (1999) appears to align with the need for this type of support for parents of children with PWS. In particular, this research indicates that parents reported a high desire for information about their children's development and the course and consequences of the syndrome. This information could likely be obtained through printed and electronic sources, such as books, pamphlets, websites, and journals. Moreover, although Turnbull, et al. (2006) suggested that primary support should come from encounters with professionals, they also recommend that families with children with PWS obtain support from clearinghouses, books, journals, and the internet for reduction of stress and improved coping.

Speaking more broadly, support through informational sources has been recommended as a means for providing information to the general population of parents of children with special developmental and health care needs. Westling (1997) surveyed parents of children with developmental disabilities, and found that parents reported wanting "much more" information on their children's syndromes and care than they had access to. Similarity, Tehee, Honan, & Hevey (2009) reported that for parents of children with autism, access to information about developmental trajectories, as well as professional community services, are needed in order to improve access to services and thus increase adaptive parent coping. Moreover, although some research indicates that generic informational support that is not tailored to the specific family can

be unhelpful, (Wodehouse & McGill, 2009,) several other studies have found that internet and printed information can play a vital role in supporting parents of children with special developmental healthcare needs (Baum, 2004; Cook, Rule, & Mariger, 2003; Seymore, Brocke, During & Poole, 1989).

Parent Satisfaction.

Not only is the availability of sources of support important, but parent satisfaction with support services is a substantial component in evaluation of the effectiveness of supports (Simeonsson, et al., 1996). For example, Mitchel & Hause-Cram (2006) reported that utilization of, and satisfaction with professional support services was correlated with lower maternal stress and depression for mothers of children with adolescents with disabilities. Hill and Rose (2009) reported that the perceived helpfulness of support and satisfaction with support was correlated with decreased parent stress for parents of adult children with intellectual disability, and that the number of support sources utilized by a family was not correlated with parent stress. In the educational domain, researchers have found that assessing parent satisfaction with services is a key method for improving services for children with developmental disabilities (Davies & Ellison, 1995; Starr, Foy, & Cramer, 2001; Starr, Foy, Cramer, & Singh, 2006). This study aimed to investigate parent satisfaction with services across educational, professional, social, and informational sources of support through: 1) assessing parent perceived helpfulness of supports, and 2) reporting on satisfaction ratings for each category of support.

Barriers to Support.

Despite the great need for school, professional, social, and informational support, barriers to support are frequently present for families with children with PWS (Freedman & Boyer, 2000; Whitman, 2006). Whitman (2006) reported that barriers include the lack of understanding of the

excessive and unique needs of PWS families by both social and professional sources of potential support. In addition, Whitman sighted barriers in the form of negatively biased views by potential supporter regarding "poor parenting" being the genesis of child problem behaviors and food-related challenges. Freedman & Boyer's (2000) found that families caring for individuals with developmental disabilities of all ages often go with unmet needs due to lack of finances, information on and awareness of services, and overly restrictive eligibility criteria. Parents also indicated difficulty with finding providers who are local and qualified, and too few providers who could address specific developmentally atypical needs knowledgeably, holistically and positively without a negative bias (Freedman & Boyer, 2000).

In addition, studies have identified some obstacles to support for parents of children with disabilities that might have implications for parents of children with PWS. Douma, Dekkar, & Koot (2006) reported that parents cited their own lack of knowledge regarding accessing services, and the desire to solve family problems without external help as barriers to support. Other barriers that have been identified include poverty, parent characteristics (e.g., depression), and limited opportunity structures where families reside (e.g., unequal access to satisfactory medical, educational, recreational, and intervention services; Deater-Deckard, 2007; Seligman & Darling, 2007). Parent perceptions of barriers to support have not yet been addressed in published research for the PWS population.

Taken together, previous research has indicated most strongly that social support might be helpful in reducing parent stress for parents of children with PWS. However, the impact of school, professional, and printed and electronic informational support had been less well researched, and therefore conclusions about helpfulness could not be made for these sources.

Through this study, parent perceptions of the availability and helpfulness of support from educational, professional, social, and informational sources were examined.

Theoretical Model

This study was based on an ecological model, where the child is embedded in nested systems that expand out form and into the child and then family (Bronfenbrenner, 1979, Seligman & Darling, 2007). The child is a system and is also a part of the family system. The family is embedded in, and interacts with lager systems, such as educational systems, professional agencies and firms, and social networks. In this model, a change in one part of a system affects subparts of a system and visa versa, creating the need for system adaption at every level. This study examined pieces of the complex relationship between the child, the child's parents, and the larger systems that encircle the child and family. More specifically, this study examined the complex relationship between child maladaptive behaviors, parenting stress, and support from educational, professional, social, and informational sources of support that interact with the family or child. See Appendix C for a graphic display of the study's theoretical model.

Summary

Because PWS is a rare disorder that presents a plethora of, behavioral challenges, as well as other medical, developmental, and mental health concerns, parents of these children are likely to experience high levels of stress (Butler et al., 2006; Hodapp, Dikens, Masino, 1997). In particular, disorders with high levels of child maladaptive behavior symptomology, like PWS, have been frequently identified as highly correlated with parent stress (Green, Furrer, McAllister, 2007; Guralnick, Hammond, Neville, Connor, 2008; Hassall, Rose, & McDonald, 2005; Hodapp, Dikens, Masino, 1997; Horton & Wallander, 2001; Mitchell & Hauser-Cram, 2008; Plant & Sanders, 2007; White & Hastings, 2004). In addition, although support from school, professional,

social, and informal sources has been suggested as essential to decreasing parenting stress for these families, the support needs of families with children with PWS are often perceived as not adequately addressed by those sources (James & Brown, 1992; Goffb, 2006; Wyatt, 2006). In fact, the Prader-Willi Syndrome Association-USA (PWSA-USA) has recently expanded their research focus, calling for proposals focused specifically on "Schools and PWS" due to the lack of research in this domain and the struggles to attain adequate school support that parents of children with PWS have reported to the PWSA-USA (Heinemann, 2008). This study attempted to address the noted gaps in research by investigating parents' perspectives of the availability and helpfulness of support from school, professional, social, and informational domains.

More specifically, this research study was designed to respond to several unanswered questions regarding support and PWS. This study sought to describe parental perceptions of received educational, professional, social, and informational support, including the sources and types of support received by parents of children with PWS. Furthermore, this study investigated the relations between perceived support, challenging child behaviors, and parent stress. It was hypothesized that parents would report social support as significantly more helpful than other types of support. In addition, it was hypothesized that all types of support would be negatively correlated with parent stress, yet the correlation between parent stress and child behavior would be positive. Overall, this study aspired to contribute to research on PWS through offering a current description of the sources of support that are perceived as available and helpful to families of children with PWS, as well as those that are not.

CHAPTER 3

Method

The main goals of this study were to examine and describe the perceived availability and helpfulness of sources from which parents of children with PWS have received support, and to examine the relations between the perceived helpfulness of sources of support, child maladaptive behavior, and parenting stress. This chapter describes methods, procedures, and the participants involved in this investigation. The following topics are discussed: participant descriptions and recruitment methods, instrument parameters, and selected test statistics. Methods were guided by three primary research questions, outlined below.

Research Questions

1. What were the frequencies by which parents of children with PWS report that support was "not needed," "not available," "not at all helpful," "sometimes helpful," "generally helpful," "very helpful," and "extremely helpful" for the following sources of support:
    a. Educational Sources (i.e., educational personnel);
    b. Professional Sources (i.e., community, non-educationally-affiliated professionals);
    c. Social Sources (i.e., family members, friends, and social acquaintances); and
    d. Informational Sources (i.e., printed and electronic informational sources)?
2. *This statement is not necessary and does not add anything to the study. It is appropriate to ask the first question without this qualifying statement.* Of the supports that were

3. reported as being utilized, did parents of children with PWS perceive some sources of support to be more helpful than others?

   Hypothesis: Based on research on support and perceptions of families with children with other developmental disabilities, it was hypothesized that social support (i.e., support from family members, friends, and social acquaintances) would be perceived as significantly more helpful than other sources of support (i.e., educational personnel; non-educational community professionals; and printed and electronic informational sources) by parents of children with PWS.

4. How are parent perceptions of the helpfulness of support by: (a) educational personnel; (b) community, non-education-affiliated, professionals; (c) social networks (e.g., family members, friends, and social acquaintances); and (d) printed and electronic informational sources related to parent stress and child maladaptive behaviors for parents of children with PWS?

   Hypothesis: Research has suggested that support is associated with lower parenting stress, and that child maladaptive behaviors are associated with higher parenting stress for families with children with developmental disabilities. It was hypothesized that, for this sample, relatively higher levels of support perceived as helpful would be predictive of lower levels of parenting stress and that child maladaptive behavior would be predictive of higher levels of parenting stress.

Participants and Recruitment

Participants

Families were recruited from 26 states for this study. Participants were 71 children (37 females and 34 males, *M* age = 10.23, age range = 3 through 19 years old) with PWS who were

enrolled in school (preschool through 12th grade), and their parents. The majority of the children were reported as attending public schools (*n* = 53; 75%), with the minority attending private schools (*n* = 12; 17%), or being homeschooled (*n* = 4; 7%). In addition, one child was reported as attending schools for those with disabilities. Reports by parents indicated the following about child PWS subtype for the sample (*n* = 56, 15 missing): 39.4% (28) Deletion-TI or II unknown; 1.4 % (1) Deletion TII; 32.39% (23) UPD; 4.2% (3) unsure if UPD or Deletion; and 1.4 % (1) "Unique Deletion." In addition, 62% (*n* = 44) of children were reported to be diagnosed with a co-morbid conditions. Frequently sited co-diagnoses included: Scoliosis, Autism Spectrum Disorders, Hypothyroidism, Sleep Apnea, Apraxia, Diabetes, Stabismus, Asthma, Hydrocephalus , and a variety of other behavioral, mood, vision, hearing, and orthopedic disorders. See Appendix E for more information on additional diagnoses.

Parent participants were between 30 and 53 years of age (65 mothers, 1 father, and 4 "both parents," *M* age = 43.19 years). One 17 year old sibling of a child with PWS helped his or her parents to fill out surveys. Sample child and parent demographics are described in Table 1. Data from participants described in Table 1 were utilized in analysis for research questions 1 and 2.

Table 1

Child and Parent Demographics (*n* =71)

Child Age (*n*=71)	*Mean* = 10.23 years[a] *Range* = 3-19 *SD* = 4.13 14.08% (10) Age 3-5 25.35% (18) Age 6-8 28.17% (20) Age 9-12 19.72% (14) Age 13-15 12.68% (9) Age 16-19
Child Gender (*n*=71)	47.9% (34) female 52.1% (37) male

Child Ethnicity (*n*=60)	91.7 % (55) Caucasian 5 % (3) Hispanic 1.7 % (1) "Bi-racial" 1.7% (1) "Filipino-American"
Child Grade Level (*n*=71)	16.90% (12) Preschool-Kindergarten 29.58% (21) 1^{st} – 3^{rd} grade 15.49% (11) 4^{th} – 5^{th} grade 23.94% (17) 6^{th} – 8^{th} grade 14.08% (10) 9^{th}-12^{th} grade
Child Abbreviated IQ (SB-Vr; *n* = 26)	*Mean = 81.64* *SD* = 19.77 *Range* = 47-121
Qualification for Special Education (*n* = 71)	30.99% (22) Other Health Impaired (OHI) 5.63% (4) Specific Learning Disability (SLD) 7.04% (5) Speech/Language Impairment (SLI) 5.63% (4) Multiple Disabilities (MU) 9.86% (7) Developmentally Delayed (DD) 21.13% (15) Answered with more than one response 12.68% (9) Missing or unclear response 2.8% (2) Not applicable
Education Plan or Program (*n*=70)	88.7% (63) IEP 2.8% (2)504 Plan 2.8% (2) General Education (Not identified under disability law) 4.23 % (3) Not applicable
Survey Respondent's Relationship to Child (*n*=71)	91.5% (65) Mother 1.4% (1) Father 5.6 % (4) Both 1.4% (1) Other
Parent Age (*n*=49)	*Mean* = 43.19 years *SD* = 6.45 *Range* = 30-53
Marital Status (*n*=65)	74.6% (53) Married 8.5% (6) Single 7% (5) Divorced 1.4% (1) Widowed
Highest Level: Mother's Education (*n*=69)	9.9% (7) High school diploma 18.3% (13) Some college to Associate's degree 35.2% (25) 4-year college degree 16.9% (12) Graduate degree
Residential Area (*n*=67)	14.1% (10) Urban 15.5% (11) Rural 64.8%(46) Suburban

[a]Mean child age at the time of SB-Vr administration sometimes differed from child age at survey completion due to some of the SB-Vr administrations occurring during the LLNP-PWS study. 7.66.

For research question 3, four participants having greater than 10% missing GAIB-PWS data were excluded from analysis. In addition, four participants who homeschooled and therefore chose "Not Needed" (NN) for the entire educational support category were also dropped from the analysis. Two participants were also dropped due to responding "NN" or "Nav" for all informational support sources. Therefore, for research question 3, a reduced sample size utilized in multiple regression analysis procedures (n = 61). In addition a subset (n = 26) of the reduced sample (n = 61) was used for analysis that included child IQ scores. All three sample groupings appear similar in composition across demographics. The reduced samples are described in Appendix D.

Parents reported that their children were educated within the following educational settings: public school (n = 53) 74.6%; private school or private preschool (n = 12) 16.9%; homeschooled (n = 4) 5.6%, and other (n = 2), 2.8%. The majority of children were identified under federal law as qualified for special education services (n = 63; 93.4%), and had an Individualized Education Plan (IEP; n = 63; 93.4 %). Within schools, the majority of children were reported as educated within a combination of separate and regular education classroom settings. Appendix G illustrates parent reported educational service delivery settings in chart form.

Procedures

Recruitment

Upon receiving IRB approval, parents of children with PWS who were between ages 3-19 and still in school (grades preschool through 12th grade) were invited to participate in the study through email or phone call recruitment procedures. Those who agreed to participate were asked

to complete three questionnaires, which took approximately 45 minutes, and mailed them back in postage paid envelopes. A subset of parents, recruited from the PWS Clinic, were also asked permission to have the Abbreviated IQ subtests (two of the 10 scales) of the Stanford Binet Intelligence Scales-5th edition (SB-Vr; Roid, 2003) administered to their children. This administration took approximately 10 minutes. In addition, SB-Vr data were available for use from the 47 potential child participants within this study's recruitment pool. These 47 children were also in the Linking Learning with Neurodevelopmental Profiles: Management Strategies for Children with Prader-Willi Syndrome (LLNF:PWS) study and were administered the SB-Vr between May 2006 and March 2008.

Recruitment involved two similar methods, depending on whether parent/child dyads participated in the LLNF:PWS study or not. (See Appendix F for Study Flow Chart). In all cases, the voluntary nature of participation was emphasized, and those in the recruitment pool were informed that neither their refusal nor consent would affect their health care at UNC Hospitals.

Mail recruitment. Parent-child dyads that participated in LLNF:PWS but who were not clinic participants or were not likely to be scheduled for their regular clinic visits during the data collection time period were contacted by email or phone to inform them about the study and the measures that were being requested. If they expressed interest, they were asked if they would be willing to have the three questionnaires mailed to them. In addition, parents were notified that they would receive feedback reports regarding their children's behavioral strengths and weaknesses as well as Target gift cards valued at $5 as gifts of appreciation for their time. (Funding for the Target gift cards was made possible by the Tom Watson Research Grant, CDL, UNC-CH). Those parents who agreed to participate were sent informational handouts that fully explained the study, the three questionnaires, and a self-addressed, stamped envelope.

Participants were informed that return of the questionnaires would constitute consent. When surveys were not returned within four or more weeks, a reminder email was sent or a phone call was made.

Clinic recruitment. Other parent/child dyads that participated in the LLNF:PWS study attended the PWS Clinic for routine health care during this study's data collection time frame. For these families, an in-person recruitment method was used. In addition, parent-child dyads who did not participate in the LLNF:PWS study but who met inclusionary and exclusionary criteria were asked to participate while at the clinic. Several of these parent-child dyads were sent a letter prior to their upcoming clinic visit explaining the study. Their package also included a parental consent and permission form and a letter with their appointment reminder information. Some families did not receive the letter (due to last minute clinic appointments, unanticipated delayed mailing, etc.). These families were approached early in the clinic day and were given an opportunity to participate. All qualifying clinic parents, both those who were in the LLNF:PWS study as well as those who were not, were asked to fill out three questionnaires. When they consented to participate (as indicated by signing the consent forms), they were given the three questionnaires, a self-addressed, and a stamped envelope in which to return the questionnaires. In addition, parents were informed that that they would receive feedback reports regarding their children's behavioral strengths and weaknesses and $5 Target gift cards at a later date. Furthermore, clinic parents whose children did not receive the SB-Vr in the LLNF:PWS study were asked for permission to administer the two subtests to their children. Children also were asked for assent prior to administration.

Once clinic parents indicated that they wanted to participate, they were given a parental consent/permission form, and provided time to carefully read it. Parents were asked if they had

any questions and efforts were taken to ensure they were fully informed. Parents who agreed to participate were asked to sign the consent/permission forms. Parents who indicated they did not have time to finish the questionnaires before leaving the clinic were given a postage-paid return envelope for returning the questionnaires.

Before administration of the SB-Vr, children were told about the study and asked to give assent; parents were present in the room during administration. For children ages 3-6 years and children who experienced difficulty with reading, the study was verbally explained and verbal assent will be sought. Children ages 7 to 17 who read well were asked to read and sign the assent form if once they agreed to participate.

To ensure confidentiality, an identification number (ID) was given to each participant, and only the ID number, first name, date of birth, and gender of the child were included on the questionnaires and test protocols. In addition, all data was kept in a locked filing cabinet in Dr. Anne Wheeler's office at the CDL, UNC-CH. Data collected from the returned questionnaires were entered into a secure database and independently verified by a second researcher.

Recruitment results. A total, 137 families were contacted between 4/27/2009 and 2/26/2010. From this pool of families, 100 parents indicated that they wanted to participate, and surveys were returned by 78 of those 100 families. Overall, 87% (n = 68) of the 78 cases were recruited from the LLNF:PWS study, 9% (n = 7) through the PWS Clinic, and 4% (n = 3) at PWS Day. Of this sample of 78, seven cases were dropped due to greater than 10% missing data on the CSS survey (n=71).

Instrumentation

Parent/Caregiver Measures

1. *The Community Support Scale* (CSS). The CSS was created to measure perceived availability and helpfulness of support for parents of children with PWS by four types of sources of support: educational (i.e. school personnel), professional (i.e., community, non-school affiliated professionals), social support (i.e., friends, family, and social acquaintances), and informational (i.e., printed and electronic information). Because a measure could not be found that addressed the perceived availability and helpfulness of support by the four named sources for this population, a new survey, the CSS, was created. (See Appendix H for CSS). The CSS was inspired by and based on two existing surveys: the *Family Support Scale* (FSS; Dunst, Jenkins, & Trivette, 1988); and the *Parent Education Perception and Satisfaction Survey- Autism Spectrum Disorder* (PEPSS-ASD; Starr, 2000). In addition, CSS items that assessed potential behavioral challenges within the school setting were inspired by and based on the *Global Assessment of Individual's Behavior–Prader-Willi Syndrome* (GAIB-PWS Tasse et al., 2002). The FSS (Dunst et al., 1988) has been used clinically and in research to identify the areas in families' support networks that need to be strengthened or accessed to better meet the families' needs (Dunst, Trivette, & Jenkins, 1984; Hassal, Rose, & McDonald, 2005). The PEPSS-ASD (Starr, 2000) has been used in research to assess the perceptions of parent of children on the autism spectrum with regard to support and services by school personnel (Starr & Foy; 2006; Starr, Foy & Cramer; 2001). The GIAB-PWS has been used clinically to describe the behaviors of individuals with PWS M. Tasse, personal communication, February 2, 2009). Permission to use these three surveys' structure and

content in the creation of the CSS was obtained from Elizabeth Starr, PhD (personal communication, August 6, 2008), Carl Dunst, PhD (personal communication, September 22, The Individuals with Disabilities Education Act, P.L. 108-446, 118 Stat. 2647 (IDEA) Part B also informed several items within the CSS. CSS items are modeled after the PEPSS-ASD (Starr, 2000), the FSS (Dunst et al., 1988), and the GAIB-PWS (Tasse et al. 2002),.2009), and Mark Tasse, PhD (personal communication, February 2, 2009). In addition, guidance from the following consultants was used: Anne Wheeler, Ph.D. (UNC-CH), Rune Simeonsson, Ph.D. (UNC-CH), Barbara Goldman, Ph.D. (UNC-CH), William Ware, Ph.D. (UNC-CH), Gregory Cizek, Ph.D. (UNC-CH), and Janalee Heinemann (Director of Research and Medical Affairs, Prader-Willi Syndrome Association-USA). See Appendix I for a detailed discussion on the CSS's creation and content, and Appendices J and K for a descriptions of the items that comprise the categories of Educational (i.e., 17 items), Professional (i.e., 33 items), Social (i.e., 13 items), and Informal (i.e., 4 items) support sources.

After obtaining IRB approval, the CSS was piloted with three families whose feedback was sought after completing the surveys. These families uniformly indicated that the measure was straightforward and appropriately addressed the support needs for families of children with PWS. Participant feedback on the CSS was consistent with pilot feedback throughout the study,

Items on the CSS that measured the perceived availability helpfulness of support sources (i.e., items 39-107) used a Likert scale that included the following response options: "not needed," "not available," "not at all helpful," "sometimes helpful," "generally helpful," "very helpful," and "extremely helpful." The five helpfulness ratings

were scored on a scale of one to five, with "not at all helpful" scored as one, and "extremely helpful" scored as five. Mean helpfulness ratings for each support source category were as follows: educational M = 3.36; professional M =3.37; social M = 2.92; and informational M =3.30. "Not needed" and "not available" responses provided categorical data and were not on the Likert scale.

Cronbach's Alpha was computed as an estimate of internal consistency (either say internal consistency or reliability but not together) reliability on the four means (n = 71, α =.675; n = 61, α =.641). This estimation of reliability was considered reasonable given the low number of items (i.e., 4 means) used in calculation. Reliability could not be computed on individual items due to the scale including qualitative and quantitative response options. "NN" and "Nav" responses were not included in analyses for research questions 2 and 3, and CSS items 72 ("Other School Personnel") and 107 ("Other") were dropped prior to analysis due to missing data > 10%.

2. *Parental Stress Index-Short Form* (PSI/SF/SF; Abidin, 1995). The PSI/SF/SF is a direct derivative of the longer 101-item Parent Stress Index. The PSI/SF/SF is a 36-item self-scoring questionnaire that yields a Total Stress score based on three subscales: Parental Distress, Parent-Child Dysfunctional Interaction, and Difficult Child. Respondents rated statements on a five-point scale ranging from "strongly disagree" (1), to "strongly agree" (5). Once administered and scored, an overall score for Total Stress and sub-scores of Parental Distress, Parent-Child Dysfunctional Interaction, and Difficult Child Items can be obtained, as well as percentiles for each scale (PSI/SF; PSI/SF/SF; Abidin, 1995). Total Stress scores are indicated and significantly elevated above the normative population at the 90th percentile and above (Abidin, 1995). In the normative population,

the Total Stress composite on the PSI/SF/SF is correlated at .94 with the Total Stress composite full-length PSI/SF. Internal reliability for the PSI/SF/SF ranges from .80 for the parent-child dysfunctional interaction to .91 for the Total Stress scale (Abidin, 1995). Test-retest reliability ranges from .68 on the Parent-child Dysfunctional Interaction to .85 on the Parental Distress scale.

This measure was normed on parents of typically developing children (Deater-Deckard, 2004), thus validity and reliability statistics have not been reported for this measure with the PWS population. However, the measure has often been utilized in studies with parents of children with behavioral challenges and developmental disabilities (Aunos, Feldman, & Goupil, 2008; Deater-Deckard, 2004; Gupta, 2007; Johnston, Hessl, Blasey, Eliez, Erba, et al., 2003; Reitman, Currier, & Stickle, 2002; Rodrigez & Murphy, 1997; Wheeler, Hatton, Reichart, & Bailey, 2007). Statistical analysis for this study included Cronbach's Alpha to determine the internal consistency reliability of the instrument with this sample of parents of children with PWS in this study. A coefficient of .956 was computed on the PSI/SF for this sample (n = 61, M = 85.38, SD = 31.08).

Child Measures

1. *Global Assessment of Individual's Behavior–Prader-Willi Syndrome* (GAIB-PWS, Tasse, Havercamp, & Mandal, 2002). Child maladaptive behavior and social competence were measured by the GAIB-PWS. The GAIB-PWS was designed as a measure where parents and caregivers rate child behaviors for individuals with PWS from age three through adult (M. Tasse, personal communication, February 2, 2009). The GAIB-PWS is a behavior rating scale that was adapted from the Nisonger Child Behavior Rating Form (CBRF), a rating scale which was normed on children and adolescents with developmental

disabilities (Aman, Tassé, Rojahn, & Hammer, 1996; M. Tasse, personal communication, February 2, 2009). The GAIB-PWS differs from the CBRF due to three modifications that were made:

> [a] items were reworded to be appropriate for adults as well as children (e.g., argues with other children was changed to argues with peers), [b] items specific to PWS were added (e.g., skin-picking, rectal digging), and [c] items tapping obsessive and compulsive behaviors were added … from the Gedye Obsessive Speech Checklist (Gedye, 1992) and the Gedye Compulsive Behavior Checklist (Gedye,1992). The Gedye scales were developed for use with developmental disabilities and [have been] widely used to measure OCD (Obsessive Compulsive Disorder) symptoms in [the PWS] population. (M. Tasse, personal communication, February 2, 2009).

This 80-item parent-report measure uses a Likert-type scale (0 through 3) and measures social competence, food and non-food related behavioral challenges, and obsessive and compulsive behaviors. All items were designed to measure specific behavioral challenges for those with PWS (M. Tasse, personal communication, February 2, 2009). The measure has been primarily used clinically and has begun to be used in research studies (Schoch, Powell, Callanan, Havercamp, & Tasse, 2006). The GAIB-PWS was chosen for this study due to its capacity to assess the complex and phenotypically specific maladaptive behavior that is unique to PWS. In this manner, the GAIB-PWS has stood in contrast to other commonly used behavioral measures, which have been designed to rate the behaviors of less atypical populations, and have not been capable of accurately capturing many of the behaviors which are characteristic to PWS

(Hodapp, Dikens, & Masino, 1997). Statistical analysis for this study included Cronbach's Alpha to determine the internal consistency reliability of this instrument with this PWS sample. A coefficient of .975 was computed for the GAIB-PWS on this sample (n = 61, M = 123.52, SD = 68.0).

Child behavior scores were calculated using the GAIB-PWS (Tasse, Havercamp, Mandal, 2002) total score, which was obtained by tallying all numerical scores other than the social competence subscale. The social competence scale was tallied using reverse scoring and was then added to the rest of the measure's overall score for maladaptive behavior. In addition, the last four questions on the GAIB-PWS (i.e., A, B, C, D), which addressed level of interference to functioning by obsessive and compulsive behaviors, were scored with "1" for a "yes" response and a "0" for "no" response.

2. *Stanford Binet Intelligence Scales-5th edition* (SB-V, Roid, 2003). A subset of child participants in this study were administered two of the 10 scales of the SB-Vr (Roid, 2003). These two scales comprised the "routing tests" of the SB-Vr, took 10 minutes to administer, and together provided an Abbreviated IQ (ABIQ) score that allowed control for child IQ in analysis.

The Stanford Binet Intelligence Scales have been known for being the most historical and widely utilized measures of intelligence still in current use (DiStephano, Dombrowski, 2006). The SB-Vr was released in 2003 (Roid, 2003), and was normed on a sample of 4,800 individuals from the general population (i.e., matching the 2000 U.S. Census), ages 2 through 85 (DiStephano, Dombrowski). Reliabilities for the 10 subtests, including the two subtests that compose the ABIQ, ranged from .84 to .89 in the norming

sample. The routing tests included one Non-verbal and one Verbal subtest; in general, the Nonverbal and the Verbal Indexes of the SB-Vr correlate with the SB-Vr FSIQ at .96 to .97 (Becker, 2003).

As noted above, PWS and Autism Spectrum Disorders (ASD) often have similar behavioral symptomology, and both have IQ as a heterogeneous feature (Butler et al., 2006; Coolican, Bryson, & Zwaigenbaum, 2008). For those on the Autism Spectrum (AS), research has indicated that other estimates of intelligence, such as the Weschler Intelligence scales- Third Edition (WISC-III; Kaufman, 1994) and the Raven's Progressive Matrices (RPMs; Raven, 1936), have underestimated IQ (Coolican, Bryson, & Zwaigenbaum, 2008). For example, for those on the AS, the WISC-III (Kaufman, 1994) short form has been indicated as accounting for only 66% of the variance of the WISC-III Full Scale IQ (FSIQ; Coolican, Bryson, & Zwaigenbaum, 2008). In contrast, Coolican, Bryson, & Zwaigenbaum, (2008) found that the SB-Vr ABIQ accounted for 89.9% of the variance for the SB-Vr FSIQ for individuals on the AS (i.e., ranging from Autism Disorder to Asperger's Disorder, and PDD-NOS). Because of the overlapping qualities of those on the AS and those with PWS, it was expected that the SB-Vr routing tests would similarly provide accurate estimates of IQ for those with PWS.

Statistical Procedures

Data Screening and Analysis

Data analysis was based on scores form the three surveys as a means to answering the given research questions. Helpfulness scores for CSS items 39-107 were tallied into means per support categories (educational, professional, social, and informational) and per each individual

support source item. See Appendix L for more information on the scoring of the CSS per support source. Raw total scores were tallied for the GAIB-PWS as a measure of child maladaptive behavior. The PSI/SF Total Stress standard score was tallied as a measure of parent stress. The SB-V ABIQ standard scores were utilized as an indicator of cognitive functioning. For all measures 100% of scores were checked for accuracy. Descriptive statistics and percentages were also calculated. Data were screened and analyzed using the statistical package, Statistical Package for the Social Sciences, for Windows, Graduate Student Version SPSS 17.0 (SPSS Inc., 2007).

Initial Screening

In preparation for implementing repeated measures ANOVA and multiple regression procedures, preliminary analyses of data were conducted to facilitate interpretable results. Data were examined for anomalies such as non-normality and outliers that might have distorted or misrepresented results. Histograms and distributions (i.e., child maladaptive behavior, IQ, age, parent stress, and the four support variables) were screened (Ware, 2008). In addition, Mahalanobis Distance (i.e., .001) was utilized to screen for univariate and multivariate outliers.

Moreover, while conducting the repeated measures ANOVA procedure, data were analyzed to ensure that sphericity was reasonable (i.e., the error structure is spherical; Howell, 2002). Screening for the lack of multicollinearity was conducted during the multiple regression procedure to ensure that results were interpretable (Ware, 2008). More specifically, the presence of multicollinearity, which would have inflated the variances of the parameter estimates, was examined through tolerance and variance inflation factors (i.e., < 0.2, > 7, respectively; Ware, 2008, W. B. Ware, personal communication, September 22, 2009).

Where significant anomalies or missing data greater than 10% per case or item were discovered, cases or items were removed (Rassler, Rubin, & Schenker, 2008). All removal or of variables or cases occurred only after consultation by a statistician (i.e., William Ware, Ph.D.), and under the consultation and supervision of committee members (i.e., Rune Simeonsson, Ph.D., Anne Wheeler, Ph.D.). For the PSI/SF and GAIB-PWS, where missing data appeared random and was less than 10% per case or item, imputation was utilized.

Participant demographics

Descriptive statistics on participant demographics were conducted on data from study enrollment forms (i.e., child race and mother's highest level of schooling), and from CSS questions 1-12 results. The CSS provided data on child age, gender, grade, genetic subtype, school and classroom setting, and medical diagnoses; and parent age, marital status, and area of habitation (Ware, 2008).

Analyses per Research Question

Statistical analyses were chosen during consultation with and at the direction of committee member, William Ware, Ph.D., (personal communication, January 28, 2009). These proposed statistical analyses are described below by research question. Appendix M presents each research question, data sources, and statistical procedures that were used to answer questions in chart format. In addition the table highlights key assumptions for the noted statistical procedures and how those assumptions were explored.

1. *What are the frequencies by which parents of children with PWS report that support is "not needed," "not available," "not at all helpful," "sometimes helpful," "generally helpful," "very helpful," and "extremely helpful" for the following sources of support:*
    a. *Educational Sources (i.e., educational personnel);*

b. *Professional Sources (i.e., community, non-educationally-affiliated professionals);*

c. *Social Sources (i.e., family members, friends, and social acquaintances); and*

d. *Informational Sources (i.e., printed and electronic informational sources)?*

To answer research question 1, support source items on the Community Support Scale (CSS) that corresponded to each support construct were computed and presented in regard to frequencies or percentages and means. (See Appendix O). Frequency distributions were calculated to ascertain the nature of support that participants perceived they received from the four categories of support. More specifically, for each of the four sources of support, the percentage of the seven possible responses (i.e., "not needed," "not available," "sometimes helpful," "generally helpful," "very helpful," and "extremely helpful") were tabulated.

2. *Of the supports that were reported as being utilized, did parents of children with PWS perceive some sources of support to be more helpful than others?*

Question 2 was answered through using the repeated measures ANOVA procedure. This statistic is often utilized to examine within group differences for a group of participants who are given the same measure repeatedly, over time. The repeated measures ANOVA can also be utilized to measure differences in participant responses to various scales at one time (Howell, 2002, p. 519; Tabachnick & Fidell, 2007).. When used in this manner, mean differences on subscales within one measure given at the same time can be explored (Tabachnick & Fidell, 2007). The repeated measures ANOVA was used in this manner to compare CSS mean perceived helpfulness scores across educational, professional, social, and informational support source categories.

Mean CSS helpfulness ratings for individual participants were calculated for each source of support for valid items, that is, those not indicated as "not needed" or "not available" by individual participants. Valid support source item scores ranged from one through five. "Not helpful at all" responses were scored as "1," "sometimes helpful" responses were scored as "2," and each increase in helpfulness on the scale was scored one ordinal unit higher than the previous, with "extremely helpful scored as "5." Where F statistics indicated significant differences between means, post hoc analyses were calculated. The Bonferonni's Method of Multiple Comparisons procedure, which examined pair-wise contrasts while controlling for Type I errors, was utilized to ascertain which means were indicated as significantly different from one another.

3. *How are parent perceptions of support by: (a) school personnel;(b) community, non-school-affiliated, professionals;(c) social networks (e.g., family members, friends, and social acquaintances); and (d) printed and electronic informational sources related to parent stress and child maladaptive behaviors for parents of children with PWS?*

To answer this question, a multiple regression procedure, was utilized to provide information on the relationship between independent (predictor) variables and the dependent (criterion) variable, parent stress. Independent variables included the 4 CSS support variables, and child maladaptive behavior, age, and IQ.

Two separate procedures were conducted. (1) The first procedure controlled for child age, and investigated parent stress as a function of the perceived helpfulness of support source categories (educational, professional, social, and informational support) and child maladaptive behavior. This analysis did not control for estimated child cognitive abilities. (2) Because estimated IQ data was gathered on only a subset of

participants, the second procedure used a reduced subsample (n = 26); the same procedures as above were conducted, with the added control variable of estimated child IQ. Both sets of results were evaluated regarding each independent variable's contribution to the prediction of parent stress (i.e. regression coefficients). Positive regression coefficients were interpreted as indicating positive relations between variables and parent stress; conversely, negative regression coefficients were interpreted as indicating negative relations. Values near "0" were interpreted as indicating the absence of relationships. It was expected that these procedures would identify which support variables best predicted decreased or decreased stress.

CHAPTER 4

Results

This chapter presents the results of statistical analyses conducted to answer the questions this study seeks to answer. Data screening results are discussed, and the results of analyses chosen for exploring each research question are provided. Results for question one provide an account of support services that are perceived as available and utilized, not needed, and not available for this sample. For research question two, supports that are reported as utilized are compared by category on perceived helpfulness. Results for question three are examined regarding how effectively support, child behavior, child age, and estimated IQ might predict parent stress.

Preliminary Analysis

Data Cleaning

Initially, data sets for 78 recruited participants were received and entered into Excel databases. All data were examined for scoring and coding accuracy; each survey was scored and entered by one party, and cross checked for accuracy of scoring and entry by another party. Subsequently, exploratory analyses were conducted to screen for anomalies and missing data. Missing data were examined via percentages by case, by total survey, and for each survey item that would be incorporated into statistical analysis. Missing data per case were most prevalent for

the CSS and GAIB-PWS surveys, which were comparatively much longer than the PSI/SF and thus more time consuming for participants to complete.

Cases with greater than 10% missing data on the CSS were removed from analysis (Langkamp, Lehman, & Lemeshow, 2010; Saunders et al., 2006). Case deletion for items missing at >10% reduced the total number of participants by seven cases for analyses pertaining to research questions one and two (n=71). For research question three, cases with an entire CSS support category rated as "NN" (n = 6) were removed from analysis. Four of these respondents described themselves as homeschoolers, and recorded "NN" for all educational support items. The other two cases responded with "NN" across the informational category. One of the two cases who indicated not needing informational support was self-described as "Hispanic" and also filled out the surveys with the help of a teenage child. Hence, a language barrier may have been a reason for responding with NN across the informational category. Additionally, cases with greater than 10% missing data on the GAIB-PWS (n = 5) were removed from analysis. These reductions resulted in a sub-sample of n = 62 which was utilized in statistical procedures for research question 3. Visual examination of missing data patterns suggested a random pattern and pages skipped on the longer surveys.

Additionally, each survey item was examined for patterns of missing data per item. Items with greater than 10% missing were removed. CSS items 72 ("Other school personnel Describe: _____," 14 missing) and 107 ("Other: _____ ," 53 missing) were removed due to > 10% of missing data. Throughout analyses, data were frequently screened to prevent errors due to mistakes in data handling (van den Broek, Cunnigham, Eeckels, Herbst, 2005).

Subsequent to case- and item-deletions noted above, imputation was utilized to fill in remaining missing data values for the GAIB-PWS and PSI/SF (Rassler, Rubin, & Schenker, 2008; Tabachnick & Fidell, 2007). Data were imputed through the SPSS Missing Values Analysis: Expectation Maximization (EM) function. The EM function estimated a variety of single imputation methods, which were processed through the EM algorithm and produced predicted values for missing items (SPSS Inc., 2007). Imputed values comprised 6.% of GAIB-PWS values and 1% of PSI/SF values Missing data for the CSS were not imputed due to the dual scaling of CSS response choices (i.e., nominal and ordinal). However, the there were few missing CSS item responses for questions used in analyses after case deletions (i.e., 1% of all responses were missing).

Preliminary Analyses

Further exploratory analyses were conducted to screen for outliers and to examine normality across variables included in repeated measures ANOVA and multiple regression procedures (i.e., helpfulness support category means; parent stress; and child age, behavior, and estimated IQ). Bivariate outliers were assessed through Mahalanobis Distance statistics (Tabanichnick & Fidell, 2007) using a threshold of $p < .001$; the presence of outliers was not indicated. Two univariate outliers were discovered via exploring boxplot output. One case contained an outlier on the child age variable (i.e., 19 years old). This family was retained in the study as the individual with PWS was residing with parents and attending public school, and therefore met the key requirements for the study's purposes. The second outlier indicated an abnormally high child maladaptive behavior score. Analyses for research question three were run with and without this case to ensure valid results. Normality was assessed through examining

histograms, and kurtosis and skewness statistics. Although descriptive statistics indicated mild kurtosis and skewness across variables, data fell within appropriate limits to this regard. Additionally, correlations between variables and were visually examined for relations that could suggest anomalies in the data. Correlations seemed to align with expectations, given current literature, and will be presented in Chapter 4, Results, and discussed in Chapter 5, Discussion and Implications.

Descriptive Statistics

Descriptive statistics for the four support helpfulness means, parent stress, and child age, maladaptive behavior, and abbreviated IQ scores are listed in Appendix N. Average perceived support helpfulness category scores ranged from 2.91 to 3.40 across all four CSS categories.

The mean PSI-SF Total Stress score was 89.34, and 54 % of participants' Total Stress scores were at the clinically significant level. PSI/SF scores indicated that 54% of the sample perceived their children with PWS as difficult, and 44% indicated experiencing dysfunctional parent-child interactions. PSI/SF scores indicated that 17% were experiencing significant parenting distress, and 7% were indicated as having exceptionally low parenting stress.

GAIB-PWS child maladaptive behavior ratings ranged from 34 to 300, with the mean at 122.78. Mean ratings for obsessive/compulsive behaviors around non-food situations (M = 14.20) were similar for obsessive/compulsive behaviors around food (M = 11.63). Scores for externalized maladaptive behaviors were the same regarding food and non-food situations. (M = 23.514; M = 23.507). Parents reported that obsessive/compulsive behaviors significantly interfered with daily activities across social, school, leisure, and home life 49.25% of the time.

Qualitative ranges had not been developed for GAIB-PWS at the time this study was conducted, so behavioral scores could not be described in terms of normed behavior severity

ratings. The GAIB-PWS scale (with reversed social competence scaling) is designed so that rating totals can range from 0 to 390. However, given that the scale measures many uniquely unusual and impairing behaviors that are often characteristic of PWS (e.g., stealing food, skin picking, obsessive and ritualistic behaviors, throwing temper tantrums, destroying property, sudden changes in mood, etc.), a mean score of 122.78 appears to indicate high maladaptive behaviors within this sample.

SB-Vr Abbreviated IQ scores (n = 26) resulted in a mean cognitive functioning estimate for this sub-sample in the Low Average range (M = 82.89) with scores ranging from the Very Low (47) range to the (121) Superior range of estimated cognitive functioning. Although IQ is a notably heterogeneous trait for the population of individuals with PWS this score was elevated in comparison to previous indications of average IQ for this population (55-70; Butler et al., 2006; Kundert, 2008; Whittington et al., 2004; Whitman & Thompson, 2006). This score appears consistent however, with approximately 1/3 or greater of the PWS population in general.

Analysis by Research Question

Subsequent to preliminary screening and cleaning procedures, statistical analyses were conducted as explicated in Chapter 3, Methods. Below, analysis findings are reported by research questions.

1. What are the frequencies by which parents of children with PWS report that support is "not needed," "not available," "not at all helpful," "sometimes helpful," "generally helpful," "very helpful," and "extremely helpful" for the following sources of support:
    a. Educational Sources (i.e., educational personnel);
    b. Professional Sources (i.e., community, non-educationally-affiliated professionals);
    c. Social Sources (i.e., family members, friends, and social acquaintances); and

d. Informational Sources (i.e., printed and electronic informational sources)?

Because data was sparse regarding perceived educational, professional, social, and written and electronic informational support for parents of children with PWS, and because this analysis is descriptive, this analysis was considered exploratory.

Mean percentages for sources of support rated on CSS as "Not Needed" ("NN") and "Not Available" ("Nav"), and those support sources that were rated as utilized (i.e., responses that fell in the helpfulness scale regarding aiding the family in raising a child with PWS) were calculated per support source category, and per individual support sources within categories (n = 71). Mean perceived helpfulness scores for support sources' helpfulness in aiding members of a family with raising a young child with PWS were also tabulated per category and per individual support source within categories. Results by category are displayed in Appendix O.

Support source categories were rated as utilized between 86% and 41% (Informational: 86%; Social: 69%; Educational: 54%; Professional 41%). Support source categories were rated as not needed between 36% and 11% (Professional: 36%; Educational: 25%; Social: 13%; Informational: 11%). Support source categories were rated as not available between 23% and 3% (Professional: 23%; Educational: 21%; Social: 18%; Informational: 3%). Mean helpfulness ratings of utilized supports ranged from 3.40 to 2.95 (Professional: 3.40; Educational: 3.38; Informational: 3.28; Social: 2.95).

Percentages of individual educational, professional, social, and informational support sources rated as not needed, not available, and utilized, and mean helpfulness ratings of utilized supports are reported in Appendi x O. Mean helpfulness ratings per individual support sources and number of respondents who reported utilizing those sources (i.e., number of respondents who rated sources on the helpfulness scale) are displayed in Appendix O.

Within the educational category, parents reported individual support sources as utilized between 87% and 7 %. (See Appendix O). The support sources that were rated as utilized by 75% of the sample or greater were: Special Education Teachers (87%); School Administrators (80%), School Speech Therapists (80%); School Nurse (76%); and Teacher Assistants (76%). Educational support sources that were rated as utilized by less 25% of the sample less were: Tutors (23%); School Social Worker (18%); and School Dietician (7%). The following educational sources were rated as most helpful by those who reported utilizing those supports: Special Education Teachers (*M* Helpfulness: 3.98; *n* = 61); Teacher Assistants (*M* Helpfulness: 3.81; *n* = 53); Tutors (*M* Helpfulness: 3.75; *n* = 16); One-on-one School Aides (*M* Helpfulness: 3.63; *n* = 27); and School Speech Therapists (*M* Helpfulness: 3.54; *n* = 56). (See Appendix O). The following educational sources were rated as least helpful by those who reported utilizing those supports: School Dietician (*M* Helpfulness: 1.80; *n* = 5); School Counselor (*M* Helpfulness: 2.52; *n* = 27); School Psychologist (*M* Helpfulness: 2.69; *n* = 34); Cafeteria Staff (*M* Helpfulness: 2.79; *n* = 42); and School Nurse (*M* Helpfulness: 2.89; *n* = 54).

Within the professional category, parents reported individual support sources as utilized between 97% and 1 %. (See Appendix O). The support sources that were rated as utilized by 75% of the sample or greater were: Pediatricians (97%); Children's Dentist (97%); Specialized Medical Personnel (97%), Foundations for PWS (85%); and Extra Curricular Activities (76%). Professional support sources that were rated as utilized by less 25% of the sample less were: Non-school-based Speech Therapists (22%); Professional Respite Care (21%), "Other" Support Groups (19%); Non-school-based Social Workers (12%); Family Therapists (11%); Clinics for those with Developmental Disabilities (10%); Developmental Therapists (8%); Neuropsychologists (7%); and Financial Counselors (1%). The following professional sources

were rated as most helpful by those who reported utilizing those supports: Non-school-based Speech Therapists (*M* Helpfulness: 4.33; *n* = 16); Early Childhood Intervention Programs (*M* Helpfulness: 4.15; *n* = 27); Prader-Willi Clinics (*M* Helpfulness: 3.92; *n* = 38); Non-school-based Physical Therapists (*M* Helpfulness: 3.91; *n* = 23); Parent Workshops/Retreats (*M* Helpfulness: 3.88; *n* = 32), Foundations/associations for PWS (*M* Helpfulness: 3.87; *n* = 64);); Non-school-based Occupational Therapists (*M* Helpfulness: 3.86; *n* = 22); and Professional Respite Care (*M* Helpfulness: 3.80; *n* = 16). (See Appendix O). The following professional sources were rated as least helpful by those who reported utilizing those supports: Departments of Social Services (*M* Helpfulness: 2.71; *n* = 17); Ministers/Religious Leaders (*M* Helpfulness: 2.46; *n* = 35); Clinics for Developmental Disabilities (*M* Helpfulness: 2.43 *n* = 7); Neuropsychologists (*M* Helpfulness: 2.20; *n* = #5; and Financial Counselor (*M* Helpfulness: 1.00; *n* = 1).

Several specialized professional support sources, (e.g., financial counselor, lawyer, hotline) were reported as not needed at high frequencies. Non-school based speech, occupational, and physical therapists were rated as not needed close to double the percentage of not available ratings. This is in contrast to the same services within the school domain, where parents reported more utilization of these supports. Overall, it appears that a high number of mental health supports across categories were rated as not needed and not available at > 60% of the sample (e.g., school-based counselor, other support group, private psychologist, respite care). In addition, traditional supports that typically address family system needs (e.g., family therapist, school- and non-school- based social worker) were indicated as utilized relatively infrequently (< 60%). Family support sources which were idiosyncratic to PWS were indicated as used at higher

frequencies than traditional family intervention supports (e.g., PWS Parent-to-parent or Family-to-family supports, Foundations for Those with PWS, PWS Clinics).

Within the social category, parents reported individual support sources as utilized between 96% and 30 %. (See Appendix O). The support sources that were rated as utilized by 75% of the sample or greater were: My Spouse/Partner (96 %); My Friends (93%), My Relatives (86%); My Children (82 %); and My Parents (79%). No social support sources that were rated as utilized by less 25% of the sample. The following social sources were rated as most helpful by those who reported utilizing those supports: My Spouse/Partner (*M* Helpfulness: 4.12; *n* = 67); My Parents (*M* Helpfulness: 3.55; *n* = 56); My Children (*M* Helpfulness: 3.38; *n* = 58); and Babysitters (*M* Helpfulness: 3.10; *n* = #42. (See Appendix O). The following social sources were rated as least helpful by those who reported utilizing those supports: Social Groups/Clubs (*M* Helpfulness: 2.38 *n* = 21); Spouse's Relatives/Kin (*M* Helpfulness: 2.25; *n* = 52); and Co-workers (*M* Helpfulness: 2.10; *n* = 31).

Within the informational category, parents reported individual support sources as utilized between 96% and 68 %. (See Appendix O). All informational support sources were rated as utilized by 75% of the sample or greater except Medical Journals (68%). Informational sources were rated between 3.49 and 3.07 on the helpfulness scale by those who reported utilizing those supports: Internet Information on PWS (*M* Helpfulness: 3.49; *n* = 66); Pamphlets on PWS (*M* Helpfulness: 3.31; *n* = 66); Books on PWS (*M* Helpfulness: 3.20; *n* = 60); and Medical Journal Articles on PWS (*M* Helpfulness: 3.07; *n* = 46).

Although barriers to support are not directly useful in answering research question one, descriptive findings are mentioned here due to the likely link between perceived barriers to support and support source items that are perceived as not available (Nav). Parents reported

barriers to accessing support services that were perceived as needed for their children with PWS and related family needs on the CSS. Participants were asked to check any of 16 barriers that they were experiencing as causing difficult regarding obtaining needed support. Out of 16 barriers possible, respondents (n = 71) reported an average of 3.51 barriers that caused difficulty in accessing the support they perceived that they needed to aid them and their families in raising children with PWS. Twenty-two participants (31.0%) cited greater than 5 barriers to adequate support. Items that were perceived as barriers with highest relative frequencies (i.e., cited as a barrier by 40% of the sample or greater) were: lack of time; lack of money; and lack of adequate understanding of PWS by local professionals in the domains of medical, school, mental health service provision. Information in chart form on perceived barriers to support sources for families in this sample is presented in Appendix O. Although analysis of parent satisfaction was not specific to research question 1, results on overall satisfaction with support across the four broad categories of support sources were tabulated for descriptive purposes. The analysis indicated that the majority of participants felt satisfaction with supports across the support source categories. Ratings of "completely satisfied" were more frequent for educational support than for the three other categories. Information in chart form on perceived satisfaction with support sources is presented in Appendix O.

Parents in this sample reported utilizing professional sources of support relatively less frequently, and informational sources of support relatively more frequently. Parent perception of helpfulness across all support was indicated in the generally helpful range. Mean helpfulness ratings per category were fairly consistent at educational M = 3.19; professional M = 3.28; social M = 2.86; and informational M = 3.26. Differentiation of ratings of helpfulness across categories was explored further through research question 2 analyses.

2. *Of the supports that were reported as being utilized, did parents of children with PWS perceive some sources of support to be more helpful than others?*

   Hypothesis: Based on research on support and perceptions of families with children with other developmental disabilities, it was hypothesized that social support (i.e., support from family members, friends, and social acquaintances) would be perceived as significantly more helpful than other sources of support (i.e., educational personnel; non-educational community professionals; and printed and electronic informational sources) by parents of children with PWS.

In order to analyze supports that participants were actually using, CSS support source items were dropped from analysis where "Not Needed" ("NN") and "Not Available" ("Nav") responses were greater than 60%. (See Appendix O). The average percent of "NN" plus "Nav" responses per support source item was approximately 50% for all of the 67 CSS support source items (45.97% educational; 59.47% professional, 31.10%; social; and 13.84% informational). After examining percentages "NN" plus "Nav" responses per support source individual items, it appeared that a cut point at greater than a 60% "NN" and "Nav" yielded the most useful data. Although this cut point was approximately 10% over the mean percentage of "NN" plus "Nav" across support sources, it allowed for support services where at least 40% of the participants reported usage to be retained (n = 29 or greater). In addition this cut point allowed for certain sources of support that were designed to support populations such as those with PWS to be retained (e.g., case manager, parent workshops and retreats, foundations that serve those with developmental disabilities, genetic counselors, paper to parent and family to family support). All together, 25 support source items were removed from analysis: six from educational support 18 from professional support and one from social support. No support source items were removed

from informational support. In total, 42 support source items were retained: 11 educational 15 professional, 12 social, and 4 informational items. The mean helpfulness rating for each support category for support sources items that were utilized by 40% or more participants (n = 65) is provided in Appendix P. These means were used in research question 2 analyses.

Also six cases were removed for research question 2 analyses (n=65) due to respondents choosing "NN" across an entire category. Four of these families reportedly homeschooled their children and choose "NN" for all education items. The other two participants chose "NN" for each informational support source item. For the subsample of homeschoolers, there appeared to be relatively similar mean helpfulness scores across the categories of professional, social, and informational support

A repeated measures analysis of variance was conducted to determine if there were significant differences between at least two of the support source category means (Tabachnick & Fidell, 2007). With this statistic, independent variables are administered simultaneously, while accounting for differences associated with cases (i.e., differences associated with individual participants are evaluated and subtracted from the error term). Bartlett's Test of Sphericity was significant, $T = 90.42$, $\chi 2\ (5) = 90.42$, $p < .001$, suggesting that the observed matrix does not have approximately equal variances and equal covariances. To avoid inflation of Type I Errors, the Huynh-Feldt epsilon correction was utilized, resulting in Huynh-Feldt $\varepsilon = .60$. Analysis of variance (ANOVA) results indicated that means were significantly different, with corrections for sphericity, $F(2,145) = 5.65$, $p = .003$. However, the effect size ($\eta^2_L = .081$) was small, indicating little strength in relationships between variables (Rosnow & Rosenthal,1996). Despite the small effect size, a straight Bonferonni method was used to examine pair-wise comparisons of the within subjects variables while correcting for the family-wise error rate. Educational and

professional mean helpfulness ratings were significantly higher than those of social support ($p < .05$). However, the small strength of effect size indicated that these results were not based on strong relationships between variables. These results are displayed in chart form in Appendix P.

Taken together, the hypothesis that social support (i.e., support from family members, friends, and social acquaintances) would be perceived as significantly more helpful than other sources of support (i.e., educational personnel; non-educational community professionals; and printed and electronic informational sources) by parents of children with PWS was not supported. For this sample ($n = 65$), social support was perceived as less helpful than both professional and educational support. Social support was not perceived as differentially helpful from informational support.

3. *How are parent perceptions of the helpfulness of support by: (a) educational personnel;(b) community, non-education-affiliated, professionals;(c) social networks (e.g., family members, friends, and social acquaintances); and (d) printed and electronic informational sources related to parent stress and child maladaptive behaviors for parents of children with PWS?*

   Hypothesis: It was hypothesized that, for this sample, relatively higher levels of support perceived as helpful would be predictive of lower levels of parenting stress and that child maladaptive behavior would be predictive of higher levels of parenting stress.

Prior to conducting regression analyses, bivariate relationships were examined across the variables of perceived parent stress and support (educational, professional, social, and informational), perceived child behavior, and child abbreviated IQ and age. Correlation coefficients were computed and the results are presented in chart from in Appendix Q. Significant relationships were found between the following variables at $p < 0.01$: child behavior

and parent stress ($r(59)= .669$; $p < .001$); social support and parent stress ($r(59)= -.374$; $p = .003$); social support and child behavior ($r(59)= -.427$; $p = .001$); social support and professional support ($r(59)= .370$; $p = .003$); educational support and professional support ($r(59)= ..621$; $p < .001$); and professional support and informational support ($r(59)= .530$; $p < .001$). In addition, significant relationships were found between the following variables at $p < 0.05$: parent stress and educational support ($r(59)= -.283$; $p = .027$); parent stress and professional support ($r(59)= -.310$; $p = .015$); social support and educational support ($r(59)= .293$; $p = .022$); and child age and abbreviated IQ score ($r(59)= -.487$; $p = .012$). Hence, at the bivariate level of analysis, perceived parent stress was significantly correlated with perceived child maladaptive behavior, and perceived helpfulness of social and professional supports for this sample.

A multiple regression analysis was conducted to evaluate how well child behavior, child age, and perceived helpfulness of educational, professional, social, and information support predicted perceived parent stress levels ($n = 61$). Variance Inflation Factors (VIF) did not indicate multicollinearity among support constructs (VIF range = 1.065 through 2.568). Analyses indicated that the predictor variables accounted for a significant amount of parent stress ($R^2 = .519$, $F(6,54) = 9.701$, $p < .001$). Only child behavior was significantly related to parent stress scores ($r = .609$, $p < .001$). These results are presented in chart form in Appendix Q.

A second multiple regression analysis was conducted to evaluate how the same independent variables predicted perceived parent stress levels when the variable of child abbreviated IQ was added to the model ($n = 26$). As with the first regression, Variance Inflation Factors (VIF) did not indicate multicollinearity among variables (VIF range = 1.513 through 2.388). For this second regression, the predictor variables accounted for a significant amount of parent stress, $R^2 = .590$, $F(7,18) = 3.693$, $p = .012$. Analyses again indicated that only child

behavior was significantly related to parent stress variance (r = .697, p = .008). Results are presented in chart form in Appendix Q. Analyses were also re-run using child age at the time of SB-Vr administration (n = 26; 61), and resulted in no change in outcomes, indicating robust results regarding child behavior accounting for a large percentage of variance in the model.

Taken together, the hypothesis that relatively higher levels of support would be predictive of lower levels of parenting stress, and that child maladaptive behavior would be predictive of higher levels of parenting stress is partially supported. With this sample, levels of support did not significantly predict parent stress, but child maladaptive behavior did. This finding was not affected by the inclusion of child abbreviated IQ.

CHAPTER 5

Discussion and Implications

This study sought to describe the sources of support that were perceived as available and helpful to a sample of parents of children with PWS. This study explored perceived support by professional, social, school personnel and informational sources for parents of children with PWS. Additionally, this study sought to explore the extent to which parents stress is predicted by perceived child maladaptive behavior and perceived educational, professional, social, and informational support.

In this chapter the findings and implications of the study are discussed with potential implications for the overall population of families with children with PWS. Child and parent characteristics are described, and their relationship with parent reported school practices for their children with PWS are discussed. Next, findings regarding each research questions are discussed, including: a) support services perceived by parents as needed, available, utilized, and helpful; b) perceived satisfaction with and barriers to supports; c) how support source categories are perceived as differentially helpful; and d) relationships between parent stress and support and child behavior. Study limitations are addressed, and overall implications and contributions of the study are proposed. The chapter concludes with suggestions for future research and clinical practice.

Sample Characteristics and Descriptives

Given the rare nature of PWS and the limited population available for research, all consenting families who met eligibility criteria were entered into the study. The participants for this study were a sample of convenience from 26 states, with the majority married, college-educated, Caucasian mothers living in suburban areas. The demographics of this study's sample were similar to those from other PWS studies (Hodapp, Dykens, & Masino, 1997; Plesa-Skwerer, Sullivan, Joffre, & Tager-Flusberg, 2004; Rosner, Hodapp, Fidler, Sagun, & Dykens, 2004). The high representation of Caucasians within this study is consistent with previous studiess indicating that PWS has been reported as occurring most often in Caucasians (Butler et al., 2006; Kundert, 2008).

The findings of this study were consistent with the previous studies that have indicated that parents with children with PWS experience high parenting stress (Hodapp et al., 1997; Whittington & Holland, 2004). The majority of parents in this sample reported clinically significant parenting stress (53.97%), which is notable when compared to the PSI norming sample where less than 10% of participant's reported clinically significant parenting stress (Abin, 1995). The parents in this study reported a 5-fold increase of stress compared the general population of parents, confirming that parents of children with PWS experience elevated parenting stress.

PWS-specific child maladaptive behaviors were reported with a high frequency (e.g., stealing food, skin picking, obsessive and ritualistic behaviors, throwing temper tantrums, destroying property). In general parents reported that obsessive and compulsive behaviors significantly interfered with child daily functioning 50% of the time. These findings are consistent with reports that children with PWS frequently demonstrate very challenging behaviors (Dykens &

Kasari, 1997; Oliver, Woodcock, & Humphreys, 2009; Rosner, Hodapp, Fidler, Sagun, & Dykens, 2004 Dykens & Kasari, 1997; Hiraiwai, Maegaki, Oka, Ohno, 2007; Kundert, 2008; University of Michigan Health Systems, 2008; Whittington & Holland, 2004). Oliver, Woodcock, and Humprhreys (2009) recently reported that many externalizing behavior problems across the sub-domains of aggression and temper outbursts might be associated with obsessive/compulsive type behaviors. Although this study did not explore relationships between these two factors, parents in this sample indicated both high obsessive/compulsive behaviors and externalizing behavioral problems.

Child abbreviated IQ for a subsample (n = 26) presented a wide range of standard scores (47 - 121),as measured by the (*Stanford Binet Intelligence Scales-5th edition* Abbreviated IQ battery (SB-V, Roid, 2003). Intelligence was a heterogeneous trait within the sample. However, the mean IQ of the subsample was higher than expected *(M* = 81.64). More specifically, compared to previous findings, the range minimum was similar, although more participants scored in the average to above average range than previously reported (Butler et al., 2006; Whittington & Holland, 2004; Whittington, Holland, Webb, Butler, Clark, et al., 2006).

Parent Reported School Services for their Children with PWS

This study provided new information via parent reported school placement and school support services for their children with PWS. Three fourths of the sample children were reported as attending public school settings (n = 53), 17% were in private schools (n = 12), and four families had chosen to home school (6%). Taken together, the majority of child participants attended public school; therefore, perceptions regarding educational support sources primarily pertained to public school personnel. Educational support services reported as provided through public and private schools were similar.

The majority of the children who attended public or private schools were identified as having an educationally relevant disability (91%) and had an IEP (94%) or 504 Plan (3%). Within public and private schools, 43% of parents reported that their children with PWS were in self-contained settings. Thus, parents reported that a large number of the children in the sample were being educated in predominately separate settings, away from typically developing peers. If parent perceptions in this regard are accurate, a high percentage of the children with PWS were being educated in what would be considered highly restrictive educational placements (McLeskey, Henry, 1999; McLesky, Tyler, & Saunders, 2004). Future research should examine educational placement of children with PWS more closely in regard to the possibility of behavioral challenges precipitating placements within restrictive settings. Eight percent of parents reported that a full time one-one-one aide who worked with their children with PWS in the school setting.

S*upport Source Use*

Children with PWS typically present with significant medical, behavioral, cognitive, educational, emotional, and developmental challenges (Butler et al., 2006). Due to the nature and breadth of the needs of children with PWS,, demands for support typically extend beyond parent capabilities to address them, apart from varied support sources. As expected, the parents within this study reported utilizing a variety of support sources cross the four categories of sources over a 6-month period. (See Appendix O).

Perceptions of *Educational Support Source*

Several educational support sources were perceived on average to be helpful and highly utilized: special education, teachers, school speech therapists, and teacher assistants. One-on-one aides and tutors were described as relatively helpful, but were utilized less frequently. School-based dieticians were rated as usually not utilized and relatively low in helpfulness even though

they could be expected to address dietary issues for children with PWS within the school setting. In addition, several other support sources that could be expected to address the behavioral, emotional, dietary and health-related needs associated with PWS were utilized by only 45% of the parents and included: school counselors, psychologists, nurses, and cafeteria staff.

Perceptions of Professional Support. Sources

Families in this study reported utilizing 41% of 33 professional support sources within the past 6 months, and that many of those support sources were extremely helpful. (See Appendix O). These findings regarding the utilization and perceived helpfulness of professional support sources are not consistent with Hodapp et al.'s (1997) study which indicated that professionals provided little support that was helpful for families in raising and caring for their children with PWS. This discrepancy could be due to differing methodological approaches, as Hodapp et al. used an opened ended format and asked participants to list "important" supports. Conversely, this study provided a comprehensive list of supports and asked for participants to response to them.

The professional sources that were perceived as most helpful were non-school-based speech therapists and early childhood intervention programs *(n* = 71). Although perceived as helpful by those who accessed this support source, private speech therapists were only utilized by 24% of the sample. This low percentage of use despite perceived helpfulness could be explained by low a perceived need for private speech therapy or barriers that prevent access. Early intervention programs, by design, would only have been utilized by parents with preschool-aged children. Other professional supports that were perceived as relatively helpful and utilized *(n* = 71) included: foundations/associations for PWS; PWS clinics; specialized medical professionals, parent workshops/retreats, parent to parent and family to family supports, and pediatricians.

Several sources of professional support that were rated as helpful by those who utilized them, but utilized relatively less than other support sources. These were professionals outside the schools and included: social workers; occupational therapists; physical therapists; and respite care providers. Several of these sources could seemingly help with family needs, such as: service provision and coordination and family functioning (i.e., social workers, Whittman, 2006); developmental challenges (i.e., occupational and physical therapy), and stress relief (i.e., respite care; Butler et al., 2006; Kundert, 2008; Deater-Deckard, 2004; Douma, Dekkar, & Koot, 2006; James & Brown, 1993). It is noteworthy that one third of the professional supports that were reported as utilized the least were rated as most helpful by those who did utilize them. Two supports that were rarely used by parents in the sample were neuropsychologists and financial counselors. Overall, professional support sources that were particularly designed for families with children with PWS (e.g., PWS clinics, PWS foundations, and family to family support) were rated as relatively more helpful than the majority of professional supports, which did not specifically address the PWS population.

Perceptions of Social Support Sources

Spouses and partners were perceived as the most helpful and most utilized social support source by parents. The following sources of support were also perceived as relatively helpful and available: parents (i.e., usually the mother's parents), children, babysitter, church members, and spouses'/partners' parents. Support sources that were rated relatively low regarding helpfulness and access included social groups and co-workers. Kin of spouses/partners were rated as relatively less helpful, despite relatively high utilization of this support source. Overall, it appears that immediate family members and participant's parents were perceived as the most helpful sources of social support.

Perceptions of Informational Support Sources

Offering generous informational support to this population has been suggested a means of supporting families with children with PWS, but previous studies have not explored the utilization of helpfulness of information supports for this population. In this study, informational support sources were utilized with high frequencies and rated as helpful overall. Books, internet sites, and pamphlets on PWS were rated as particularly helpful. This finding is consistent with van den Borne, van Hooren, van Gestel, Rienmeijier, Fryns, et al.'s (1999) research indicated that parents of children with PWS reported high desire for access to informational support sources.

Barriers.

Parents reported an average of 3.51 barriers in accessing the support they perceived as needed in raising their children with PWS (n = 71). Twenty-two participants (31%) cited between 5 to 9 barriers to acquiring adequate support. Items that were perceived as barriers with highest relative frequencies (i.e., cited as a barrier by 40% of the sample or greater) were: lack of time; lack of money; and lack of adequate understanding of PWS by local professionals in the domains of medical, school, mental health service provision. These findings were consistent with the findings of other researchers (Freedman & Boyer, 2000; Whitman, 2006). In particular, Whitman noted that the lack of understanding of PWS by support providers as a substantial barrier to adequate support and services for families with children with PWS.

Satisfaction.

Results indicated that participants felt more satisfaction than dissatisfaction with support, and ratings of "completely satisfied" were more frequent for educational support than for the three other categories. Mitchel & Hause-Cram (2006) reported that utilization of, and satisfaction

with professional support services was correlated with lower maternal stress and depression for mothers of children with adolescents with disabilities. Within this study, the complex relationships between parenting stress and satisfaction with, the perceived helpfulness of, and access to support sources and were not explored. Future research could investigate these relationships.

Cross Categorical Themes

In addition, to the above noted findings, certain themes arose across support categories in the domains of mental health, behavioral, and developmental supports. Ratings of parent stress in this study were high. Use of hotlines by 24% of the participants (n = 16) within the past 3 to 6 months may also have been indicative of high rates of stress. However, a large number of mental health support sources designed to address family functioning were frequently not rated as utilized: social workers, counselors, psychologists, psychiatrists, respite care, and family therapists..Conversely, use of supports unique to PWS was indicated as utilized at relatively higher frequencies compared to traditional family intervention supports: foundations for those with PWS, PWS Clinics, and parent-to-parent and family-to-family supports. Turnbull, Erwin, & Soodak (2006) suggested that support for the families of children with PWS is necessary, and they noted that PWS-specific parent-to-parent and family-to-family support sources could be utilized in this regard. These findings indicate that PWS specific support sources are helpful for families in raising children with PWS.

Parents frequently endorsed the fact that their children with PWS had been diagnosed with speech and motor delays. (See Appendix E). This finding was consistent with what is typically found in the general population of those with PWS (University of Michigan, 2008). However, parents rated support by non-school-based speech, occupational, and physical

therapists, who typically address speech and motor functioning delays in the professional domain, as not needed ("NN") close to double the percentage of not available ("Nav"). Moreover, developmental therapists, whose role is often to facilitate speech and motor development, were endorsed as utilized at a relatively low frequency. In contrast, parents endorsed the support of school speech, occupational, and physical therapists in schools at higher frequencies (i.e.,). Hence, developmental supports in the schools were utilized more often than those within the professional setting. Given the high number of developmental, medical and behavioral needs of the children with PWS in the study, the elevated stress level of their parents, and the financial barriers to accessing supports, families in this study may have attempted to meet their children's needs within the convenience of a school setting wherever possible. This finding is similar to Haveman, van Berkum, Reijnders, & Heller's (1997) finding that parents of children with developmental disabilities rated support services accessed within educational settings as more essential supports for their children's development than those found within other settings.

Statistical Comparison of Support Source Categories

Statistical analyses for Research Question 2 (n=65) did not indicate significant differences between mean helpfulness of support source categories for supports that were utilized (i.e., by > 40% of the sample), and the hypothesis that social support sources would be perceived as more helpful than other support sources was not supported. This finding is in contrast to findings from populations with children with other developmental and behavioral disabilities where social supports have been reported as more helpful that other types of support sources (Deater-Deckard, 2004; Guralnick, Hammond, Neville, & Connor, 2008; Hodapp, Dykens, & Masino, 1997; Plant & Sanders, 2007; Seligman & Darling, 2007; Suarez, & Baker, 1997; White & Hastings, 2004). Undifferentiated helpfulness ratings across educational, professional, social,

and informational support sources could be explained by the significant needs that children with PWS have across behavioral, health, medical, developmental, academic, social, and emotional domains (Bulter et al., 2006; Kundert, 2008). It is possible the breadth and intensity of child needs necessitates using support sources across the four domains (Wigren & Hansen, 2003; Whitman; 2006; Wyatt, 2006). Also, the inclusion of proximal (i.e., spouses/partners) and distal (i.e., co-workers) social support sources may have impacted results. The lack of understanding of PWS by professionals and educators was indicated as a barrier to accessing support, and a similar barrier may have been present in social acquaintances outside of proximal social support sources (e.g., spouse, children, and parents). Hence, individuals within families' social networks who had less exposure to, and investment in the child with PWS may have provided less helpful support due to a lack of understanding and empathy PWS and its impact. Seligman & Darling, (2007) indicted that support by family members might be most important for families with children with developmental disabilities. The impact of support provided by distal versus proximal social support on parent stress should be an area of future research for families of children with PWS.

Parent Stress as a Function of Child Behavior and Support

An analysis (n = 61) of bivariate relationships between the variables of parent stress, child behavior, age, and abbreviated IQ (n = 26), and the perceived helpfulness of educational, professional, social, and informational support sources indicated significant correlations between several variables ($p < 0.01$ or $p < 0.05$). As expected, child behavior was positively correlated with parent stress. In addition, consistent with findings for other disabilities(Guralnick, Hammond, Neville, & Connor, 2008; Hodapp, Dykens, & Masino, 1997; Plant & Sanders, 2007; Seligman & Darling, 2007; Suarez, & Baker, 1997), perceived helpfulness of social support was negatively correlated with parent stress. Social support was also positively correlated with child

behavior. Perceived helpfulness of professional support followed the same correlation pattern as social support. Professional and educational support's significant negative correlation with parent stress is partially consistent with James and Brown's findings (1993), indicating correlations between parent stress and access to community and social services, such as respite care and special education support. Consistent with Hodapp et al.'s findings (1997), IQ was not correlated with parent stress. In addition, perceived helpfulness of professional support was significantly correlated with the perceived helpfulness of social, educational support, and informational support.

Despite the significant bivariate correlations between parent stress and support source variables, support from the four support source categories did not account for significant variance when child behavior was present as another predictor variable in regression analyses. At the multivariate level, the variance accounted for by child maladaptive behavior was significantly greater than the variance accounted for by the four support source categories. These finding support Hodapp et al.'s (1997) work, which indicated child behavior is a strong predictor of parent stress for parents of children with PWS.

Through the view point of this study's theoretical framework, findings indicated that children and family systems within the study had been interacting with a large number of educational, professional, social, and informational support sources within the community state, and national systems that surrounded them (Bronfenbrenner, 1979, Seligman & Darling, 2007). However, the evidence that access to those systems of support (i.e., four categories of support sources) might predict decreased parenting stress was not found when child maladaptive behavior was present. (See Appendix O).

Limitations

When exploring implications of this study, it is important to consider certain limitations. Similar to other studies within the domain of PWS, this study used a sample of convenience (Dykens & Kasari, 1997, Hodapp et al., 1997 Oliver, Woodcock, & Humphreys, 2009). Hence, the results could be confounded by parent characteristics associated with those who might participate within this type of study. The sample demographics are similar to other studies as well, but caution should be used when generalizing results across the characteristics of racial diversity, marital status, and parent education (Butler et al., 2006, Dykens & Kasari, 1997, Hodapp et al., 1997 Oliver, Woodcock, & Humphreys, 2009).

Another limitation of the study might be the potential impact of missing data on results. Dropping cases can skew data, especially if missing data were not by random occurrence (Rassler, Rubin, & Schenker, 2008). As noted previously, an analysis of missing data indicated predominately random patterns. It does not appear that missing data were due to individuals opting not to respond due to a personal characteristic. Imputation was utilized for two surveys after case-wise deletions were made. Although original data is preferred, imputation is preferable to case-wise deletions when attempting to minimize bias (Rassler, Rubin, & Schenker, 2008).

Neither validity statistics (e.g., internal and construct validity) nor reliability statistics (e.g., test-retest and internal consistency, and reliability) were available prior to the study for the Community Support Scale (CSS) and the Global Assessment of Individual Behavior- PWS (GAIB-PWS; Tasse et al., 2002). However, post data collection analyses contributed to the reliability data for the GAIB-PWS, and indicated acceptable internal consistency. The CSS presented challenges in determining internal reliability due to scaling. Moreover, sequence effects may have impacted statistical outcomes on perceived helpfulness of support sources for

Research Questions 2 and 3, as items on the CSS survey were not counter-balanced (i.e., were not provided in randomized order to participant). Parent stress associated with time of diagnosis could have been a confound for a few parents within this study. However, the majority of children in the study (87%) had participated in the Linking Learning with Neurodevelopmental Profiles: Management Strategies for Children with Prader-Willi Syndrome (LLNF:PWS) two years ago, and therefore were not newly diagnosed.

Implications and Future Directions

Given the significant likelihood of elevated parent stress for parents of children with PWS, surprisingly little research had been conducted, prior to this study, on the helpfulness of sources of support and the complex relationship between parenting stress, child behavior, and support.. An important finding of this study is that, despite usage of support sources across the four categories of support, analyses did not indicate that utilized support contributed significantly to a decrease in parent perceived stress. The prediction of elevated parent stress by child maladaptive behavior significantly surpassed the prediction lower parent stress by the four support source categories. In regard to decreasing parenting stress that is predicted by child maladaptive behaviors, parents' usage of perceived and available support sources has not been effective. Hence, with regard reducing parent stress, this study indicates the need for a re-evaluation of support sources across educational, professional, social, and informational sources to inform recommendations for families of children with PWS (Wigren & Hansen, 2003; Whitman; 2006; Wyatt, 2006). New information provided by this study on barriers to sources of support could also be considered within any such re-evaluation.

This study contributes knowledge regarding perceptions of helpfulness, and perceived utilization of educational, professional, social, and informational support by parents of children

with PWS. Statistical comparisons of means did not indicate that parents perceived significant differences in helpfulness for the four categories of sources of support. However, descriptive results of mean helpfulness ratings for individual support sources (i.e., within categories) indicated that some individual sources of support might be perceived as more helpful than others. Future research could statistically explore this possibility. Furthermore, this study provides a current picture of support that is perceived as not needed, not available, and helpful by parents of children with PWS which could be cautiously generalized to those with similar demographic characteristics.

Implications for Research. Social, educational, professional and informational support sources have been identified as effective agents in lessening parenting stress (Deater-Deckard, 2004; Grant & Whittell, 2000; Haveman et al., 1997; Ogletree, Fischer, & Shultz, 1999). Yet, the findings of this study indicate the need for further research to clarify if and how support sources might decrease parent stress for parents of children with PWS, given frequently elevated maladaptive child behaviors. Future research could explore which individual supports might significantly decrease parent stress as well as reduce child maladaptive behavior. Research should also evaluate the quality of information parents are accessing from the internet and the extent to which it is a source of support. This study could be replicated, with the aim of examining the role of parent educational level and race on perceptions of support. The study could also be re-designed in order to explore differences in the support source utilization and perceived helpfulness of sources of support across broader demographic parameters. At the time of this study, access to accurate PWS subtype data was not available. However, future research could explore the impact of genetic subtype on child behavior and parent stress. Barriers to, and satisfaction with support sources could be further explored in relationship to perceived

helpfulness of support sources and parent stress. In addition, support seeking as a parent characteristic could be explored in relation to parent stress and child behavior outcomes.

Implications for Clinical Practice. The findings of this study may have implications for those who provide interventions for children with PWS and their families. (See Appendix S). Parents seemed to perceive individual support sources that were specifically designed for to PWS as helpful more frequently than those that were less specific to PWS. They also indicated that lack of knowledge about PWS by professionals and educators was often a barrier to accessing support. These findings could indicate the need for greater access to supports that are specific to PWS (i.e., PWS clinics, PWS foundations, and PWS parent-to-parent and family-to-family supports).

Better dispersion of knowledge across educational and professional support sources could help with improving the quantity and quality of support that families receive. Results indicated more use of proximal supports sources (i.e., those found in the everyday school and home environments) rather than specialists. In particular, proximal social and educational support sources (i.e., spouses, teachers) were indicated as highly utilized. However, proximal supports are typically not equipped to address the complex behavioral profile and family impact of PWS. Results indicated less use of specialists who should be equipped to intervene with child behavioral and parent stress challenges (e.g., school and non-school counselors, therapists, psychologists). Parents indicated that a primary barrier to accessing support by professionals in school and community settings was perceived lack of understanding of PWS. It is possible that the perceived lack of understanding by specialists facilities the overuse of proximal, everyday supports and the underuse of specialists. The overuse of proximal sources and under use of specialists could have negative long-term effects. Families might over-tax basic everyday

systems of support (e.g., immediate family members, regular and special education teachers, and babysitters), resulting in a decrease of quality and quantity of support from proximal sources, and thus experience further unmet family needs. Dispersion of information regarding PWS to specialists could remove barriers to accessing these support sources.

Whereas families appeared to be relying heavily on certain proximal social and educational support sources, certain professional sources of support that were accessed infrequently should be used more often. Because private speech, occupational, and physical therapists were rated as helpful, yet not often utilized, PWS clinics should consider including these disciplines in regular treatment of children with PWS. Community supports that are often paid for by state and federal funds, such as community case managers and developmental therapist were infrequently used by this sample. PWS advocacy organizations might attempt to: 1) provide parents with knowledge on how to obtain these supports; and 2) provide easily accessible training for these paraprofessionals (e.g., via the internet). In addition, dieticians were not rated as highly utilized, nor as relatively helpful by this sample. PWS foundations and clinics might consider easily assessable training materials for school-based and private dieticians who have clients with PWS as well.

Within the domain of educational support sources for children with PWS, two supports were indicated as utilized at low frequencies but rated as highly helpful: one-on-one aides and tutors. Educational and community support providers might seek to facilitate greater access to these support sources through advocacy. In addition, several individuals who provide support within educational settings and who were often utilized but rated as less helpful could be better trained to respond to the needs of children with PWS in the school setting: school administrators, nurses, cafeteria staff, and psychologists, counselors.

These findings indicate that child maladaptive behavior significantly predicts parent stress for this population. Hence, support providers should be aware of the possible impact of PWS that extends beyond the child to the parents (i.e., stress), and facilitate interventions for all impacted parties. In addition, given the reported perception of spouses and proximal social supports as particularly helpful, support providers that address family needs should be aware of family composition. Those without adequate family or proximal social support may benefit from compensatory supports (Seligman & Darling, 2007). For example, parent-to-parent programs and support groups should provide compensatory proximal social support. Furthermore, given the large number of individual supports that families indicated utilizing over a span of six months, providers might consider better coordinating services in a manner that could lessen the burden of finding and accessing sources of support.

Appendix A
Etiology and Physical, Medical, and Developmental Outcomes

This appendix reviews, in detail, the physical and medical symptomology of PWS, as well as the genetic etiology and the clinical and genetic elements of diagnosis.

Genetics

PWS is caused by a malfunction on the 15q11-q13 chromosomal region leading to the loss of expression of paternal genes (Buiting & Horsthemke, 2006). This region is under the control of an imprinting center. Although the process of imprinting is not completely understood, it is known that parent-of-origin specific DNA methylation (a modification that occurs in DNA as for regulation of imprinted genes) plays a central role in imprinting. In non-PWS individuals, the 15q11-q13 region contains a group of genes that are maternally imprinted, and only the paternally inherited copy is active (Whittington & Holland, 2004). More specifically, the majority of genes are methylated (and silenced) on the maternal chromosome and expressed solely from the unmethylated paternal allele (Buiting & Horsthemke, 2006). PWS is caused by one of several possible malfunctions in this process, each of which results in the absence of the paternal contribution to the 15q11-q13 region (Buiting & Horsthemke, 2006).

Within in the syndrome, there are 3 subtypes that are based on malfunctions within pre-expression processes: 1) deletions and translocation; 2) maternal uniparental disomy (UPD); and 3) imprinting defects (Buiting & Horsthemke, 2006). Paternally originated deletions of the 15q11-q13 region (including the imprinting and many non-imprinted genes) account for approximately 70% of PWS cases, and occurs randomly in 1 out of 10,000 live births (Buiting & Horsthemke, 2006;Whittington & Holland, 2004). Researchers suggest that breakages occur because the region is bounded by unstable DNA repeat sequences, which allow for rare cases of sporadic deletion and even more rare translocations (Whittington & Holland, 2004). At the molecular level, researchers have identified two types of deletions, Type I (TI) and Type II (TII), which involve deletions at different break points on the gene sequence. TI deletions occur in 30% to 40% of deletion cases, whereas TII deletions occur in 60% to 70% of cases. Research has indicated that the PWS critical region has three breakpoints (BP); TI deletions involve longer breaks, spanning from BP1 to BP3. TII deletions, on the other hand, involve breaks spanning only in the BP2 through BP3 area (Zarcone et al. 2007). Hence, more genetic material is lost with TI as opposed to TII, leading to relatively more severe impairment in functioning for those with TI deletions (Zarcone et al., 2007). Chromosome 15 translocations, where part of one chromosome is broken off and attached to a different chromosome, occurs very rarely (5% of cases; Whittington & Holland, 2004); hence, most PWS cases are caused by deletions (Buiting & Horsthemke, 2006).

UPD accounts for approximately 25% of cases of PWS, and is caused by an individual receiving two maternal sets of chromosome 15 due to a maternal meiosis malfunction. Hence the individual inherits three sets of 15q11-q13 genes, one paternal and two maternal. Here, the duplicate maternal sets of genes cause paternally expressed genes to be silenced. The maternal genes cannot compensate for the lack of paternal genes and PWS occurs.

Lastly, imprinting causes approximately 1% of cases of PWS. Within this subtype, an individual inherits an original set of intact paternal and maternal chromosome 15 genes, but the paternal chromosome carries a maternal imprint, causing loss of paternal expression. For parents of children with PWS, the recurrence rate for new conceptions is less than 1%, except for imprinting errors where the recurrence rate can be as high as 50% (Buiting & Horsthemke, 2006).

Diagnosis

PWS occurs across all ethnicities and races, but is reported as occurring most often in Caucasians (Butler et al., 2006; Kundert, 2008). Just a decade ago, diagnosis did not occur until more severe symptoms, such as obesity, were observed (Butler et al., 2006; McCandless & Cassidy, 2006); however, the current diagnostic statistics show a trend towards earlier diagnosis (Kundert, 2008; Wigren and Hansen, 2003). Current data indicate that 29% of diagnoses are made by the age of one, 53% are made by the age five, and 89% are diagnosed by the age of seven (Kundert, 2008; Wigren and Hansen, 2003).

Diagnosis begins with a hypothesis of PWS, based on evidence from observation symptomology congruent with clinical criteria (McCandless & Cassidy, 2006). After adequate clinical evidence prompts further investigation, diagnostic genetic testing is administered and these results confirm the diagnosis (McCandless & Cassidy, 2006). Although not diagnostically conclusive, clinical criteria are useful for early identification and in preventing costly genetic testing where superfluous (McCandless & Cassidy, 2006).

The majority of clinical criteria are based on patterns of development that occur across two distinct phases. Phase I begins *in utero* with decreased fetal movement, and a high rate of malposition (McCandless & Cassidy,

2006). Then, as infants, those with PWS display extreme hypotonia, and often have weak suck reflex, leading to breastfeeding failure and poor intake from bottle-feeding (Butler et al., 2006). With frequent failure in weight gain, many of these infants require long-term (i.e., 6 to 9 months) feeding tube interventions (Butler et al., 2006). Usually, Phase I begins to ebb around the end of the first year, and is typically followed by a short period of resolution of feeding difficulties and slow upward development (i.e., two to three years; McCandless & Cassidy, 2006)). This period is called the "honeymoon period" because it precedes Phase II where hyperphagia and new behavioral challenges begin (McCandless & Cassidy, 2006).

In Phase II, those with PWS have excessive appetite as well as a lack of hunger satiation, even after eating large amounts of food (McCandless & Cassidy, 2006). Behavioral problems begin during this time as well (McCandless & Cassidy, 2006). Clinically relevant symptoms in Phase II include obsession with food and non-food objects, foraging and hording of food, intense temper tantrums and episodes of loss of emotional control, repetitive questioning, difficulty with transitions, and excessive need for routine and structure (McCandless & Cassidy, 2006). In addition, unless intense food-related interventions are utilized (i.e., heavily monitored diet and locking up off-diet food), obesity frequently develops (Butler et al, 2006; Kundert, 2008). Beyond obesity, physical criteria can include hypogondalism, short statue, mildly abnormal facial characteristics, and small hands and feet (McCandless & Cassidy, 2006). In addition to the noted behavioral and physical symptomology, global delays in development and learning difficulties are also indicative of PWS (Butler et al, 2006). Once a health care provider observes clinical features consistent with Phase I or Phase II symptomology, genetic testing is typically recommended for diagnostic clarity (McCandless & Cassidy, 2006).

Diagnostic evaluation for PWS typically occurs a step-wise manner. First, PWS is confirmed using a methylation-sensitive DNA method (e.g., PCR-based or Southern blotting; McCandless & Cassidy, 2006). However, occasionally false negative results occur (McCandless & Cassidy, 2006); hence, repeat testing is recommended when clinical evidence and genetic results are not consistent (McCandless & Cassidy, 2006). Once abnormal methylation that is consistent with the PWS genotype is confirmed, a fluorescence in situ hybridization (FISH) analysis is performed to assess for the subtypes of deletion or UPD; if neither are found, imprinting is assumed by default (McCandless & Cassidy, 2006). In addition, chromosome analysis is occasionally ordered to rule out translocation (McCandless & Cassidy, 2006).

Physical Development and Characteristics

The physical characteristics of PWS include obesity, growth retardation, fair skin, small hands and feet, short stature, and dysmorphic facial features (i.e., narrow face, almond shaped eyes, and small mouth with thin upper lip and down-turned corners; Kundert, 2008; State & Dykens, 2000; University of Michigan Health Systems, 2008). Other features include hypotonia (weak muscles and low muscle tone), fatigue, and strabismus (i.e., cross eyes; Kundert, 2008; State & Dykens, 2000; University of Michigan Health Systems, 2008). Furthermore, hypoganadalism (i.e., low levels of sex hormones) and delayed or incomplete gonadal maturation are almost universally present (Crinò et al., 2003; Kundert, 2008). In addition to these features, physical symptomology is present throughout the developmental stages of prenatal growth, infancy, childhood, and adolescence through adulthood.

Prenatal development and infancy. In utero, the most common physical characteristics are decreased fetal movement and frequent breech presentation (25%; Butler et al., 2006). In addition, retarded prenatal growth is common, and low birth weight is seen in 30% of newborns (Butler et al., 2006). Also higher rates of asphyxia and non-term (early or late) deliveries are common (Butler et al., 2006; University of Michigan Health Systems, 2008). Moreover, little interest in food coupled with feeding difficulties frequently result in failure to thrive and feeding tube interventions for infants. (Whittington &Holland, 2004). In addition, severe hypotonia persists into early childhood, and overall motor milestones are delayed (University of Michigan Health Systems, 2008). Other symptoms that frequently begin in infancy are: hypogenitalism (i.e., partial or complete failure of the genitalia to develop); and in males, hypogandalism (i.e., decreased production of gonadal hormones), and cryptorchidism (i.e., failure of descent of one or both of the testes into the scrotum; Butler et al., 2006).

Childhood. Typically, between 18 months to 2 years of age, feeding problems resolve, and by age five or six, hyperphagia (i.e., insatiable appetite) develops. If intense weight and diet management strategies are not stringently employed, severe obesity can occur as early as two to four years old. Additionally, endocrine abnormalities (e.g., hypothyroidism) are also common, and can exacerbate weight problems and decrease energy and activity levels (Butler et al., 2006). Often, growth hormone is used to improve both energy level and short stature (Butler et al., 2006).

In addition, developmental milestones are typically delayed throughout childhood (Butler et al., 2006). For example, the mean age of walking is 27 months, and speech disarticulation is common, in part due to hypotonia in

oral musculature (Atkin & Lorch, 2007). Other problems include dental problems (e.g., enamel hypoplasia, dental carries), which occur due to soft tooth enamel, thick saliva, teeth grinding, and poor oral hygiene (University of Michigan Health Systems, 2008). Vision difficulties (e.g., nystagmus, strabismus, myopia, decreased visual sharpness, and impaired stereoscopic vision) also frequently are present (Butler et al., 2006). Furthermore, many children (e.g., close to 50% in a sample of 497) develop delayed bone age, scoliosis, or seizures (Butler et al., 2006; Whittington & Holland, 2004). High pain thresholds, bed-wetting, sleep difficulties and sleep apnea also present frequently in childhood (University of Michigan Health Systems, 2008).

Adolescence and adulthood. Most of the afore mentioned physical challenges continue into young adulthood, with severity varying based on individual differences, as well as type, intensity, and length of early interventions. In addition, adolescents and adults with PWS also face new challenges in physical development; for example, scoliosis occurs at a high frequency, and bone problems such as osteoporosis can occur earlier than in the typical population (University of Michigan Health Systems, 2008). Moreover, delayed or absent puberty frequently occurs, which causes adolescents and adults to appear awkwardly immature when compared to their same age peers, (Butler et al., 2006). Lack of a normal puberty is caused, in part, by low gonadotropin production as well as various other endocrine malfunctions (Butler et al., 2006). Small male genitalia are sometimes treated with testosterone, and gonadotropin treatment is utilized for cryptorchidism (Butler et al., 2006). For females, menarche often is absent (70%; Butler et al., 2006), and pregnancy is extremely rare (i.e., 2 known cases; Butler et al., 2006). Various hormone treatments are somewhat helpful in achieving additional growth, but stature, size of hands and feet, and sexual maturity rarely reach typical levels (Butler et al., 2006).

Obesity is a primary health concern for adolescents and adults with PWS. In fact, PWS is noted as the most common form of obesity rooted in a genetic cause (Butler et al., 2006). These problems are so pervasive that researchers have described PWS as a physiologically driven "eating disorder," where individuals remain on one of the two extremes of the eating continuum (Greenswag & Alexander, 2006). After remaining at one end of the continuum during infancy, where poor ability to ingest nourishment is life threatening (Butler et al., 2006), individuals quickly shift to intense hyperphagia as young children; hyperphagia is almost universally in full bloom by adolescence (Kundert, 2008). By if uncontrolled, hyperphagia can lead to obesity levels that are life threatening, in childhood, adolescence, and adulthood (e.g., one third weigh 200% more than ideal body weight; Butler et al., 2006). Weight problems are exacerbated by short stature, low metabolic rate, impaired emesis (i.e., ability to vomit), and decreased need for calories (i.e., needing 40% to 70% fewer calories than typical peers; Butler et al., 2006; University of Michigan Health Systems, 2008). Obesity is more likely to be life-threatening if trained caregivers are not in place to manage overeating behaviors (Butler et al., 2006).

Serious medical problems follow obesity for these individuals. Complications such as high blood pressure, diabetes, cellulitis, hypoventilation, and chronic venus insufficiency (leading to ulcers and sores on legs and feet) often develop (University of Michigan Health Systems, 2008). Moreover, heart failure, hypertension, thrombophlebitis (vein inflammation related to blood clots), chronic leg edema, orthopedic difficulties, and Type 2 diabetes mellitus occur frequently (Butler et al., 20006; University of Michigan Health Systems, 2008). In addition, skin ulcers, sleep apnea impaired respiratory function, and various endocrine disturbances are a few of the other obesity-related problems that can manifest (Butler et al., 20006; University of Michigan Health Systems). However if obesity and overeating are controlled (i.e., forced exercise and extreme measures to monitor weight, including locking up all unauthorized food and food like-items), few serious health issue manifest, and life expectancy can be similar to those with mild intellectual disabilities (Butler et al., 2006).

Neurological characteristics. Research studies have begun to identify various neurological differences in those with PWS, many of which are hypothesized as facilitating hyperphagia and stunted physical development (Kundert, 2008). For instance, positron emission tomography (PET) and magnetic resonance imaging (MRI) research have indicated that in those with PWS, the hypothalamus, which is the cerebral domain that controls sensations of hunger and satiation, malfunctions (Crinò et al., 2003). This malfunction is thought to contribute to hyperphagia and obesity (Kundert, 2008). In addition, Whitman and Jackson (2006) note that the hypothalamus affects sensitivity to pain, impacts memory, and regulates breathing and temperature, day and night cycles, and emotions. All of these physiological symptoms malfunction in PWS (Whitman & Jackson, 2006). Furthermore, structural MRI and neuroanatomical autopsy studies indicate that the paraventircal nucleus, which is involved in appetite, is smaller in size and has fewer oxytocin-expressing neurons in the PWS brain (Butler et al, 2006). In addition, those with PWS have a 30% reduction in growth hormone-releasing hormone (GHRH) neurons in the arcuate nucleus, a brain area that is involved in appetite (Kundert, 2008; Swab, 1997). Similarly, Sharpio et al. (2005) found satiety dysfunction within the central nervous system (i.e., within the insular, ventromedial prefrontal cortex, and nucleus accumbens) in individuals with PWS. Other abnormalities indicated by fMRI research include

ventriculomegaly (enlarged ventricles), sylvian fissure polymicrogyria (i.e. excessive small folds on surface of brain), decreased brain stem size, decreased volume in the parietal-occipital lobe, and incomplete insular closure, all of which are thought to adversely affect development (Miller et al., 2007; Kundert, 2008).

Overall, medical-care burdens, diet and weight monitoring, and coping with developmental challenges are likely to place heavy demands on parents of children with PWS. Medical and physical management of children with PWS typically requires inordinate investments of time, finances, and emotional resources. Yet, PWS symptomology and its impact on caregivers spans beyond the physical and medical domain into every area of development, including cognitive development.

Appendix B
Cognitive, Academic, and Adaptive Characteristics of PWS

Cognitive functioning.

Generally children and adolescents with PWS exhibit lower intellectual functioning than their same-aged, typically-developing peers (Butler et al., 2006). Studies consistently point to IQ scores averaging in the mildly impaired range (55-70), or 40 points below the typically developing population (Butler et al., 2006; Whittington & Holland, 2004; Whittington, Holland, Webb, Butler, Clark, et al., 2006). However, these statistics fail to demonstrate the heterogeneous nature of cognitive functioning for this population (Butler et al., 2006). Generally speaking, approximately one third present with standard IQ scores within the 70 to 100 range, approximately two-thirds present with scores in the 50 to 70 range; and roughly 5% are indicated as functioning in the severe and profound range (Butler et al., 2006; Kundert, 2008; Whittington et al., 2004; Whitman & Thompson, 2006). In addition, IQ scores are known to decline over the lifespan, indicating a slowing of the cognitive growth curve over time (Jauregi et al., 2007; Whittington et al., 2004). Studies have been inconclusive regarding body mass index's (BMI) relationship to IQ, with some studies indicating relatively higher IQ correlating with average weight, and others failing to demonstrate these findings (Butler, 2006; Jauregi, Arias, Vegas, Alén, Martinez, et al., 2007).

Research is beginning to indicate cognitive strengths and weaknesses for those with PWS; however, to be considered conclusive, many findings would need to be replicated. Butler et al., (2006) and Dykens, (2002) note strengths in recognizing, evaluating, and integrating figures on a spatial plane, and in working with puzzles; and similarly, Whittington et al. (2004) describe relative strengths in visual, spatial, and long-term memory skills. Conversely, Whittington et al. (2004) describe weaknesses such as impaired social cognition, cognitive inflexibility, literal mindedness, and weaknesses in sequential processing and short-term memory. Moreover, Conner et al. (2000) was able to replicate findings that indicate that long-term memory is stronger than short-term memory for individuals with PWS. Moreover, although various research findings have indicated better visual processing than auditory processing, a study of academic achievement failed to produce this finding in a small sample of adults with PWS (Conners, Rosenquist, Atwell, & Klinger, 2000).

Cognitive abilities and adaptive skills are usually consistent; however daily living skills are typically higher relative to social skills for those with PWS (Kundert, 2008). Koenig, Klin, & Schulz (2004) report significantly lower performance on tasks of social attribution using visual prompts when compared with IQ-matched controls, yet comparable performance with those diagnosed with pervasive developmental delays. Another study found differences regarding social skills for those with UPD and deletion subtypes, with more impaired social skills within the UPD subtype (Milner, Craig, Thompson, Veltman, Thomas, et al., 2005).

An additional study which investigated the neurocognitive functioning of 16 adults with PWS found deficits in executive frontal cognitive processes, including deficits in global and selective attention, general executive organization and planning, visuo-perceptual organization, and sequential processing (i.e., receiving, storing, processing and using information in an orderly way; Jauregi, Arias, Vegas, Alén, Martinez, et al., 2007). However, with repetition of items, initial sequential processing deficits improved. In addition, research on performance on simultaneous processing tasks (i.e., task that required encoding information into groups and categories in memory) has indicated strength in this domain, with scores aligning with the normative population (Jauregi et al., 2007). Similar to Jauregi et al.'s findings (2007), Wigren & Hansen (2005) illustrated that those with PWS have impairment of executive functioning that is consistent with Attention Deficit Hyperactivity Disorder (ADHD).

Academic functioning.

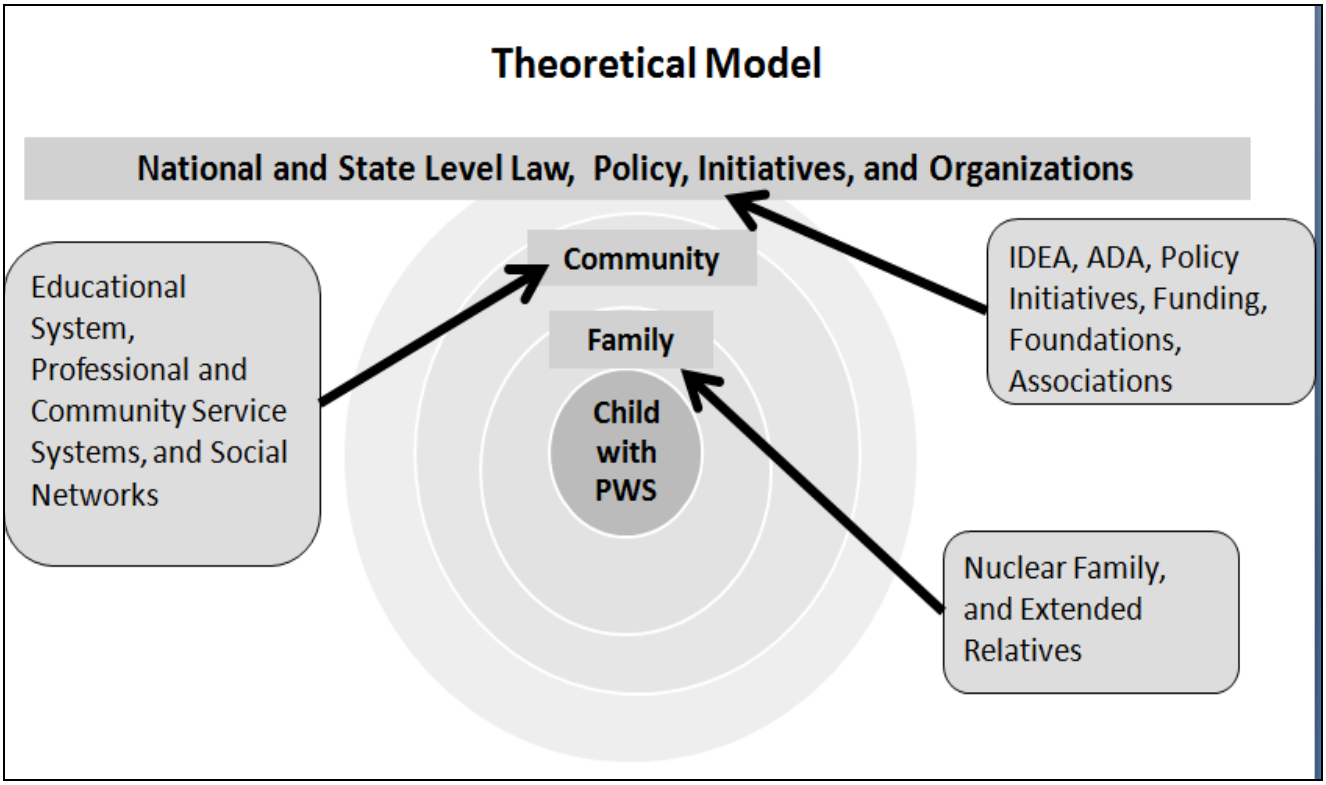
Given broad cognitive and overall developmental deficits, it is predictable that academic functioning is impaired for those with PWS. For example, studies have found discrepancies between IQ and achievement scores that reflect significantly lower academic scores (i.e., over one standard deviation) compared to indicated cognitive abilities (i.e. abilities indicated by IQ scores; Whittington et al., 2004). Downward discrepancies from IQ to achievement scores, referred to as "underachievement," have been identified for both general achievement variables as well as for specific academic areas (i.e., arithmetic, tasks involving social cognition or auditory processing, and academic skills that use short-term memory; Kundert, 2008; Whittington et al., 2004). Along with organic learning disabilities, interference by food-related and behavioral problems has been implicated as facilitating underachievement (Kundert, 2008). Research has also indicated that time spent in special schools (i.e., schools allocated for educating children with behavior problems) is correlated with underachievement for those with PWS (Whittington et al., 2004). Children with PWS are often placed in special schools due to behavioral problems; however, Whittington et al. (2004) speculate that within these schools, educational needs are masked by poor social

skills and maladaptive behaviors. Because true abilities are masked, exposure to adequate and ability-based curriculum and instruction might not occur.

Individuals with PWS also demonstrate relative strengths and weaknesses in specific academic skill domains. For example children with PWS typically exhibit relative strengths in reading and spelling when compared to non-PWS controls (Whittington & Holland, 2004), and relatively strong reading skills are especially prevalent for those with the UPD subtype (Whittington et al., 2004). In addition, some academic material can be challenging for students with PWS, who have been noted as demonstrating greater abilities in comprehending concrete material than abstract material (Whittington & Holland, 2004). Moreover, difficulties mastering multiplication facts and calculation are common (Kundert, 2008), and arithmetic scores are typically lower than their peers, and even more so for those who score in the impaired range of the IQ continuum (Whittington et al., 2004).

In summary, those with PWS have impaired intellectual and social-cognitive capacities and adaptive skills, as well as delayed academic achievement. Consequently, meeting the academic needs of these children usually necessitates significant school, caregiver, and professional academic intervention. In addition, if partial autonomy in late adolescence and adulthood is to be attained, significant support in the domain of adaptive skills and self-determination would be required (James & Brown, 1993).

Appendix C
Theoretical Model



Appendix D
Child and Parent Demographic Subsamples (n = 61; n = 26*)*

For Research Question 3.a. (*n* = 61)		For Research Question 3.b. (*n* =26)	
Child Age (*n* = 61)	*Mean* = 10.21 years *Range* = 3-16 *SD* = 4.23	Child Age (*n* = 26)	*Mean* = 9.40 years[a] *Range* = 3-16 *SD* = 4.05
Child Gender (*n* = 61)	53% (32) female 48% (29) male	Child Gender (*n* = 26)	54% (14) female 46% (12) male
Child Ethnicity (*n* = 50)	96% (48) Caucasian 4% (2) Hispanic	Child Ethnicity (*n* = 24)	100 % (23) Caucasian
Child Grade (*n* = 61)	18% (11) Preschool-K 28% (17) 1^{st} – 3^{rd} grade 17% (10) 4^{th} – 5^{th} grade 23% (14) 6^{th} – 8^{th} grade 15% (9) 9^{th}-12^{th} grade	Child Grade (*n* = 26)	19% (5) Preschool-K 37% (10) 1^{st} – 3^{rd} grade 15% (4) 4^{th} – 5^{th} grade 15% (4) 6^{th} – 8^{th} grade 11% (3) 9^{th}-12^{th} grade
Child Behavior Total (GAIB-PWS) [b]	*Mean* = 123.52 *SD* = 68.00 *Range* = 34-300	Child Behavior Total (GAIB-PWS)	*Mean* = 110.08 *SD* = 68.33 *Range* = 34-300
Respondent's Relationship to Child	92% (56) Mother 2% (1) Father 7% (4) Both	Respondent's Relationship to Child	89% (23) Mother 11% (3) Both
Parent Age (*n* = 44)	*Mean* = 44.0 years *SD* = 6.35 *Range* = 30-53	Parent Age (*n* = 20)	*Mean* = 43.57years *SD* = 6.45 *Range* = 31-53
Mother's Education (*n* = 52)	10% (5) HS Diploma 27% (14) Some College 44% (23) 4-year College 17% (9) Graduate Degree	Mother's Education (*n* = 23)	4% (1) HS Diploma 26% (6) Some College 35% (8) 4-year College 35% (8) Graduate Degree
Parent Total Stress Score (PSI/SF) [b]	*Mean* = 85.38 *SD* = 31.08 *Range* = 36-161	Parent Total Stress Score (PSI/SF)	*Mean* = 89.34 *SD* = 26.67 *Range* = 36-161

[a]Mean child age at the time of SB-Vr administration sometimes differed from child age at survey completion due to some of the SB-Vr administrations occurring during the LLNP-PWS study. 7.66.
[b]Unlike the descriptive data for the full sample (n = 71), this table includes descriptions of child behavior and parent stress, using imputed values for missing values (<10% per case).

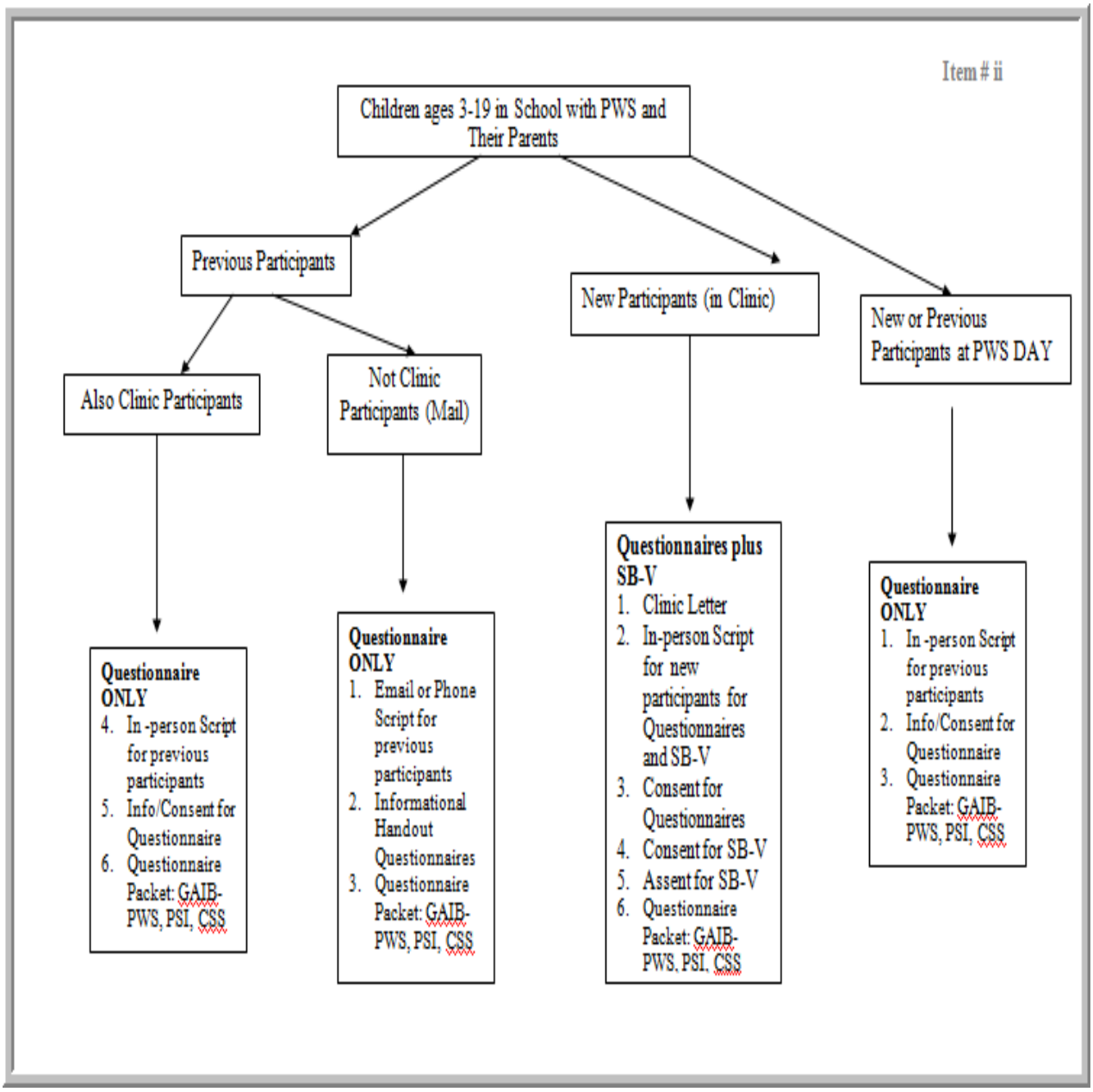
Appendix E
Reported PWS Subtype and Additional Child Diagnoses

	PWS Subtype							
	Deletion TI	Deletion TII	Deletion Unknown	Unique Deletion[a]	UPD	Unsure	No Response	Additional Diagnosis[b]
n	0	1	23	1	18	3	15	35
%	0	1.6	37.7	1.6	29.5	4.9	24.6	57.4

[a] Written in response: "Unique deletion, not TI or TII."

[b]Written in responses were: Scoliosis (11), Autism Spectrum Disorders (5), Hypothyroidism (9), Sleep Apnea (4), Apraxia (3), Diabetes (3), ADHD (3), Stabismus (2), Asthma (2), Hydrocephalus (2), Oppositional Defiant Disorder (2), Sensory Integration Disorder (2), Dyspraxia, Heart Murmur, Developmental Delays, Learning Disabilities, Speech Delays, Hearing Impairment, Ocular Albinism, Nystagmos, Vascular Problems, Astigmatism, Short Chain Amino Acid Deficiency, Central Adrenal Insufficiency, Prolapsed Rectum, Orthopedic Disorders, Behavior Disorder, Tic Disorder, Anxiety Disorder, Premature birth , Kidney Disorders, Osteochondromia, Blepharospasm, Allergies, Congenital Muscular Dystrophy, Oppositional Defiant Disorder, Mood Disorder, and Class III malocclusion.

Appendix F
Study Flow Chart



Appendix G
Parent Reported Within School Educational Service Delivery Settings (n= 71)

School Setting	*n*	*P*
F/T Regular Education Setting	1	1.40
Regular Education Setting w/ 1-on-1 Aid	5	7.00
Combination of Separate and Regular Education Settings	26	36.62
F/T Separate Setting	18	25.35
Separate School for those with Disabilities	9	12.68
Home School	4	5.63
Other[a]	8	11.27

[a] "Other" descriptions: F/T Sep Setting w/ P/T 1-on-1 Aid; Reverse Mainstream, Regular education classroom with pull out speech services.

Appendix H
Community Support Scale

Child's date of birth:_____ Today's Date_____ Identification #:_____

Community Support Scale

Modified by Barbara Lowe-Greenlee, B.A., Anne Wheeler, Ph.D, and Rune Simeonsson, Ph.D., from the Family Support Scale by Carl J. Dunst, Vicki Jenkins & Carol M. Trivette, and the Parent Education Perception and Satisfaction Survey-Autism Spectrum Disorder (PEPSS-ASD), ©Elizabeth Starr, Ph.D.

This survey is appropriate for families of children with Prader-Willi Syndrome ages 3-17.

Thank you again for your helpful input. I will start by asking you questions about your child with Prader-Willi syndrome, your family, and your child's school setting.

1. Who is completing this survey?

❑ MOTHER	❑ FATHER	❑ BOTH	❑ Other: _____

2. What is the gender of your child with Prader-Willi Syndrome (PWS)?

❑ MALE	❑ FEMALE

3. What is the age of your child? _____YEARS _____MONTHS OLD

4. Where do you and your child live? (Check one box)

❑ IN A RURAL AREA	❑ IN AN URBAN AREA	❑ IN A SUBURBAN AREA

5. Has your child had genetic testing to identify the subtype of PWS s/he has?

❑ YES	❑ NO

5.a. (If yes) Indicate the genetic type of PWS that your child has: (Check one box)

❑ DELETION-TYPE I	❑ UPD
❑ DELETION-TYPE II	❑ IMPRINTING ERROR
❑ DELETION-TYPE UNKNOWN	❑ UNSURE

6. Has your child received any diagnosis in addition to Prader-Willi syndrome or have any other medical problem(s)?

❑ YES	❑ NO

6.a. (If yes) What is the diagnosis and/or other medical problem(s)?

_____

7. What type of school does your child attend? (Check one box)

❑ PUBLIC SCHOOL	❑ PRIVATE *PRESCHOOL*
❑ PRIVATE SCHOOL (elementary, middle, or high school)*	❑ HOME SCHOOL
	❑ OTHER:_____
	❑ OTHER:_____

7.a. (If private school*) Was this school selected primarily because it is designed for children with disabilities? (Check one box)

❑ YES	❑ NO

Thank you for filling out and returning this survey.

Appendix H
Community Support Scale

Child's date of birth:_____ Today's Date_____ Identification #:_____

12. Does your child have an education team that assists in making decisions regarding your child's education?

❑ YES	❑ NO

13.a. (If no) Please explain why

_____

12.b. (If yes) Please indicate, by checking all that apply, who make up your child's team. **Check all applicable boxes.**

❑ PARENT (YOU)	❑ OCCUPATIONAL THERAPIST
❑ REGULAR EDUCATION TEACHER	❑ DIETICIAN
❑ SPECIAL EDUCATION TEACHER	❑ SCHOOL NURSE
❑ TEACHER ASSISTANT	❑ SPECIAL EDUCATION FACILITATOR
❑ PRINCIPAL	❑ ONE-ON-ONE CALSSROOM AIDE
❑ ASSISTANT PRINCIPAL	❑ SPECIALIST (Describe)_____
❑ SCHOOL PSYCHOLOGIST	❑ CONSULTANT (Describe)_____
❑ SPEECH PATHOLOGIST	❑ ADVOCATE (Describe)_____
❑ PHYSICAL THERPAIST	❑ OTHER (specify)_____

For the next set of questions, I would like to know about your child's symptoms.

For the following statements, please circle "No" or "Yes"		YES	NO
13.	Due to health concerns, my child is on a special food plan.	Yes	No
14.	Due to health concerns my child has is on a special exercise plan.	Yes	No
15.	My child sometimes struggles with worries and anxiety.	Yes	No
16.	My child sometimes shows a lot of sadness or moodiness.	Yes	No
17.	My child sometimes becomes very angry or frustrated.	Yes	No
18.	My child sometimes performs behaviors that might seem unusual (for example, any of the following: skin picking, pulling out hair, hording things, talking to imaginary friend, hearing sounds/seeing things that are not real, etc.).	Yes	No
19.	My child has some obsessive and compulsive behaviors (repetitive talking, behaviors, rituals, etc.).	Yes	No
20.	My child has a hard time with following rules.	Yes	No

Thank you for filling out and returning this survey. 3

Appendix H
Community Support Scale

Child's date of birth:_____ Today's Date_____ Identification #:_____

For the next set of questions, I would like to know about your family's experiences with your child's school. Please circle the best answer from the following:

Strongly Disagree	.....	1
Mostly Disagree	.....	2
Barely Disagree	.....	3
Barely Agree	.....	4
Mostly Agree	.....	5
Strongly Agree	.....	6
Not Applicable	.....	n/a

Please circle the best answer based on your child's current school experience.		→						n/a
21.	My child's ability to get food at school is closely watched so that my child follows his/her special food plan.	1	2	3	4	5	6	n/a
22.	My child is given enough opportunities to exercise during school hours.	1	2	3	4	5	6	n/a
23.	My child's teacher is good at helping my child cope with worried and anxious feelings.	1	2	3	4	5	6	n/a
24.	My child's teacher is good at helping my child cope with sadness and moodiness.	1	2	3	4	5	6	n/a
25.	My child's teacher is good at helping my child to calm down when very angry or frustrated.	1	2	3	4	5	6	n/a
26.	My child's teacher good at dealing with my child's behaviors that might seem unusual (any behavior similar to: skin picking, pulling out hair, hording things, talking to imaginary friend, hearing sounds/seeing things that are not real, etc.).	1	2	3	4	5	6	n/a
27.	My child's teacher is good at helping my child handle obsessive and compulsive behaviors (repetitive talking, behaviors, rituals, etc.).	1	2	3	4	5	6	n/a
28.	My child's teacher is good at handling my child's difficulties with following the rules.	1	2	3	4	5	6	n/a
29.	My child's daily classroom routine is predictable.	1	2	3	4	5	6	
30.	My child's teacher prepares him/her enough for changes in classroom routines.	1	2	3	4	5	6	
31.	My child's teacher makes sure that my child is included in activities with classmates.	1	2	3	4	5	6	
32.	My child's teacher makes sure that my child is treated well by classmates.	1	2	3	4	5	6	
33.	My child's teacher uses a reward system for good behavior that **works for my child.**	1	2	3	4	5	6	
34.	I feel my child's educational team understands my child's needs.	1	2	3	4	5	6	n/a
35.	My child's education team members **do not** make me feel that I am to blame for my child's difficult behaviors.	1	2	3	4	5	6	n/a
36.	I feel that my child's educational team values my comments, suggestions, and concerns regarding my child.	1	2	3	4	5	6	n/a
37.	My child's educational team members communicate what my child does well in addition to problems that arise.	1	2	3	4	5	6	n/a
38.	My goals and objectives for my child are included in his/her individualized education plan.	1	2	3	4	5	6	n/a

Thank you for filling out and returning this survey.

Appendix H
Community Support Scale

Child's date of birth:_____ Today's Date_____ Identification #:_____

Listed below are people and groups that are helpful to members of a family raising a young child with Prader-Willi Syndrome. This questionnaire asks you to indicate how helpful each source is to your family. Please circle the response that best describes how helpful the below sources have been to your family for the **past 3 to 6 months**. CIRCLE ONLY ONE RESPONSE (NN, NAv, 1, 2, 3, 4, or 5).

For each item:

- If you have used the particular source of help listed, circle **1, 2, 3, 4, or 5** based on how helpful you found that source of help.
- If you wish you had access to a particular source of help, but it has not been available to your family or you have never heard of it before now, circle the **NAv** (Not Available) response.
- If you believe your family has not needed a source of support during this period, despite whether or not it has been available, circle the **NN** (Not Needed) response.

How helpful has each of the following been to you in terms of raising your child with Prader-Willi Syndrome:	Not Needed	Not Avail-able	Not at All Helpful	Some-times Helpful	Generally Helpful	Very Helpful	Extremely Helpful
39. My parents	NN	NAv	1	2	3	4	5
40. My spouse's or partner's parents	NN	NAv	1	2	3	4	5
41. My relatives/kin	NN	NAv	1	2	3	4	5
42. My spouse's or partner's relatives/kin	NN	NAv	1	2	3	4	5
43. Spouse or partner	NN	NAv	1	2	3	4	5
44. My friends	NN	NAv	1	2	3	4	5
45. My spouse's or partner's friends	NN	NAv	1	2	3	4	5
46. My own children	NN	NAv	1	2	3	4	5
47. Other parents	NN	NAv	1	2	3	4	5
48. Co-workers	NN	NAv	1	2	3	4	5
49. Babysitter(s)	NN	NAv	1	2	3	4	5
50. Parent support group	NN	NAv	1	2	3	4	5
51. Other support group Describe: _____	NN	NAv	1	2	3	4	5
52. Social groups/clubs	NN	NAv	1	2	3	4	5
53. Minister or religious leader	NN	NAv	1	2	3	4	5
54. Church members	NN	NAv	1	2	3	4	5
55. My physician	NN	NAv	1	2	3	4	5
56. My child's pediatrician	NN	NAv	1	2	3	4	5
57. Specialized medical professionals (orthopedist, surgeon, etc.) Describe:_____	NN	NAv	1	2	3	4	5
58. My child's dentist	NN	NAv	1	2	3	4	5
59. My child's summer camp or retreat staff	NN	NAv	1	2	3	4	5
60. Early childhood intervention program	NN	NAv	1	2	3	4	5
61. Regular education teacher(s)	NN	NAv	1	2	3	4	5
62. Special education teacher(s)	NN	NAv	1	2	3	4	5
63. Teacher assistant(s)	NN	NAv	1	2	3	4	5

Thank you for filling out and returning this survey.

Appendix H
Community Support Scale

Child's date of birth:_____ Today's Date_____ Identification #:_____

How helpful has each of the following been to you in terms of raising your child with Prader-Willi Syndrome:	Not Needed	Not Available	Not at All Helpful	Sometimes Helpful	Generally Helpful	Very Helpful	Extremely Helpful
Special education facilitator	NN	NAv	1	2	3	4	5
64. School nurse	NN	NAv	1	2	3	4	5
65. One-on-one school aide	NN	NAv	1	2	3	4	5
66. Tutor(s)	NN	NAv	1	2	3	4	5
67. School psychologist	NN	NAv	1	2	3	4	5
68. School counselor	NN	NAv	1	2	3	4	5
69. School administration (principals, superintendents)	NN	NAv	1	2	3	4	5
70. Cafeteria staff	NN	NAv	1	2	3	4	5
71. Bus driver	NN	NAv	1	2	3	4	5
72. Other school personnel Describe: _____	NN	NAv	1	2	3	4	5
73. Private psychologist	NN	NAv	1	2	3	4	5
74. Family therapist	NN	NAv	1	2	3	4	5
75. Neuropsychologist	NN	NAv	1	2	3	4	5
76. Psychiatrist	NN	NAv	1	2	3	4	5
77. School-based dietician	NN	NAv	1	2	3	4	5
78. Non-school-based dietician	NN	NAv	1	2	3	4	5
79. School-based social worker	NN	NAv	1	2	3	4	5
80. Non-school-based social worker	NN	NAv	1	2	3	4	5
81. School-based occupational therapist	NN	NAv	1	2	3	4	5
82. Non-school-based occupational therapist	NN	NAv	1	2	3	4	5
83. School-based speech therapist	NN	NAv	1	2	3	4	5
84. Non-school-based speech therapist	NN	NAv	1	2	3	4	5
85. School-based physical therapist	NN	NAv	1	2	3	4	5
86. Non-school-based physical therapist	NN	NAv	1	2	3	4	5
87. Case manager	NN	NAv	1	2	3	4	5
88. Professional respite care for my child	NN	NAv	1	2	3	4	5
89. Financial counselor	NN	NAv	1	2	3	4	5
90. Developmental therapist	NN	NAv	1	2	3	4	5
91. Department of social services	NN	NAv	1	2	3	4	5
92. Prader-Willi clinic	NN	NAv	1	2	3	4	5
93. Clinic for those with a any developmental disability	NN	NAv	1	2	3	4	5
94. Foundations or associations that serve families with children with PWS	NN	NAv	1	2	3	4	5

Thank you for filling out and returning this survey. 6

Appendix H
Community Support Scale

Child's date of birth:_____ Today's Date_____ Identification #:_____

How helpful has each of the following been to you in terms of raising your child with Prader-Willi Syndrome:	Not Needed	Not Available	Not at All Helpful	Sometimes Helpful	Generally Helpful	Very Helpful	Extremely Helpful
95. Foundations or associations that serve families with children with any developmental disability	NN	NAv	1	2	3	4	5
96. Hotline	NN	NAv	1	2	3	4	5
97. Books on PWS	NN	NAv	1	2	3	4	5
98. Medical journals	NN	NAv	1	2	3	4	5
100. Internet information on PWS	NN	NAv	1	2	3	4	5
101. Pamphlets on PWS	NN	NAv	1	2	3	4	5
102. Parent workshops or retreats. Describe: _____	NN	NAv	1	2	3	4	5
103. Lawyer/legal advocate	NN	NAv	1	2	3	4	5
104. Genetic counselor	NN	NAv	1	2	3	4	5
105. Parent-to-parent or family-to-family support*	NN	NAv	1	2	3	4	5
106. Extra curricular activities	NN	NAv	1	2	3	4	5
107. Other: _____	NN	NAv	1	2	3	4	5

* Parents or families with children with special needs are matched by professionals in order to support one another.

108. Please check all of the following that makes it hard for you to get the support you need for your child with PWS and your family.

- ☐ Not enough time
- ☐ Not enough money
- ☐ Poor health insurance
- ☐ Lack of childcare
- ☐ No or poor transportation
- ☐ Too few knowledgeable school staff who understand my child's PWS-related needs at my child's school
- ☐ Lack of agencies with the right resources to help me with my child with PWS in my area (for example, not enough knowledgeable staff, funds, literature, etc.)
- ☐ Lack of mental health personnel who understand PWS in my area (private psychologists, social workers, psychiatrists, etc.)
- ☐ Lack of medical personnel who understand PWS in my area (nurses, doctors, etc.)
- ☐ Feeling that our family should handle things without outside help
- ☐ Lack of computer/internet access
- ☐ Difficulties due to my own disability
- ☐ People seem prejudiced against helping my child/family due to my child having PWS.
- ☐ I speak a different language and there are no interpreters
- ☐ I don't understand where to look for help or information
- ☐ Other (Specify):_____
- ☐ Other (Specify):_____

Thank you for filling out and returning this survey. 7

Appendix H
Community Support Scale

Child's date of birth:_____ Today's Date_____ Identification #:_____

For the following statements please CIRCLE the best NUMBER that indicates your level of satisfaction, with 1 being "NOT SATISFIED" being and 4 being "COMPLETELY SATISFIED."

	Overall Satisfaction	Not Satisfied	Mildly Satisfied	Fairly Satisfied	Completely Satisfied
109.	Please rate your satisfaction with the support your family and your child receive from the **school personnel** in the school where your child attends school.	1	2	3	4
110.	Please rate your satisfaction with the support your family and your child receive from **professionals outside of the school setting** (that is, medical, mental health, religious, agency, clinic professionals, etc.).	1	2	3	4
111.	Please rate your satisfaction with the support your family and your child receive from **family members, friends, and social acquaintances.**	1	2	3	4
112.	Please rate your satisfaction with the support your family and your child receive from **printed informational sources** (that is books, internet, etc.)	1	2	3	4

Thank you for filling out and returning this survey.

Appendix I
Creation and Content of the Community Support Scale

The CSS was inspired by the PEPSS-ASD (Starr, 2000) in regard to: demographic, educational, and medical information; methods for surveying hetero- and homogeneous symptomology; and, methodology for surveying parent satisfaction. The first 12 questions of the CSS asked for basic demographic information (e.g., who is completing the survey, child age, etc.), general and special educational information (e.g., child grade, IEP team members, type of school and classroom setting), and general medical information (e.g., subtype of PWS, other medical problems). These questions were modeled after questions within the PEPSS-ASD (Starr, 2000) that requested similar information (i.e., PEPSS-ASD questions 72-85; 90; 93; 100). Also, because the PEPSS-ASD (Starr, 2000) asked for more detailed (e.g., information about siblings) and sensitive (e.g., annual income, parent level of education) demographic information than was needed for the purposes of this study, many PEPSS-ASD items (1,-2, 72-76, 78, 86-87, 89, 94, 96-106) were omitted from the CSS (R. J. Simeonsson, personal communication, September 22, 2008). These omissions were also performed to decrease the burden of time for parents who would be filling out the surveys.

The PEPSS-ASD (Starr, 2000) also inspired CSS methods for surveying parent perceptions of children's educational needs while accounting for a span of within disability differences. On the PEPSS-ASD (Starr, 2000), parents of children on the autism spectrum (AS) were asked what school supports their children need based on unique AS symptomology. Then, parents were asked to respond to separate questions that assessed their perceptions of school support regarding their children's unique support needs (PEPSS-ASD; Starr, 2000; questions 4-33). For example, PEPSS-ASD question 12 examines parents' beliefs that their children need visual schedules at school, and question 13 asked if the school provides their children with visual schedules. This method allowed for investigating parent perceptions of support that was tailored to their children's unique needs, as visual schedules might not have been perceived as necessary for all children on the AS.

Similar to individuals on the autism spectrum, those with PWS have presented with heterogeneous behavioral symptomology and support needs. Because PWS has shared this diversity of phenotype with autism, Starr's (2000) two-question methodology seemed appropriate for determining parent perceptions of school support for the heterogeneous behavioral challenges and support needs for children with PWS. The CSS utilized this method in questions 13-28, but with modification in wording so that the questions were tailored to the needs of children with PWS, as identified by Tasse et al. (2002).

Items 29-38 of the CSS addressed the more homogeneously experienced behavioral and learning challenges that children with PWS typically have in the classroom (Goff, 2006). Therefore, these questions did not ask for parent opinion regarding whether or not certain supports were needed, but rather assumed those support needs. For example, in CSS question 29, parents were given the statement "my child's classroom schedule is predictable," and they were asked to rate their level of agreement with the statement. Due to problems with behavioral and cognitive shift in PWS, it was assumed that a predictable schedule would have been needed for any child with PWS (Whittington et al., 2004). Therefore, parent opinion as to whether or not a predictable schedule has been needed was not requested in the CSS.

Items 109-112 of the CSS assessed parent satisfaction with support services. These five items were modeled after question 68 of the PEPSS-ASD (Starr, 2000), which asked, "please rate your satisfaction with your child's education." In the creation of the CSS, Starr's (2000) question was expanded to capture parents' satisfaction with support across each of the four sources of support with which this study is concerned: school personnel; community (non-school-affiliated) professionals; social networks; and printed and electronic information.

Many questions on the CSS used scaling that was modified from the PEPSS-ASD (Starr, 2000). For example, CSS items that rated satisfaction with support (109-112) used a Likert scale of 1-4, by which parents rated their state of satisfaction from "not satisfied" (1) through "completely satisfied" (4). This CSS scale was similar to Starr's (2000) 5-point scale used for addressing satisfaction question (PEPSS-ASD question 68); however, a 4-point scale was chosen for the CSS in order to force a non-neutral choice (Fink, 2009). In addition, the PEPSS-ASD (Starr, 2000) used a 5-point scale ("strongly agree," "mostly agree," "mostly disagree," "strongly disagree," "unable to comment") for items (7, 10, 12, 15, 21, 26) that assessed parent perceptions of child support needs for heterogeneous AS traits. Starr (2000) used the same 5-point scale for items (8, 11, 13, 16, 22, 27) that assessed parent perceptions of school service provision designed to meet those needs. Conversely, the CSS used a "yes" and "no" format for items (13-20) that assessed parent perceptions of child support needs for heterogeneous PWS traits (e.g., "Due to health concerns, my child is on a special food plan;" Circle "yes" or no"). This change was made to simplify response choices to CSS items 13-20 (Fink, 2009). Also in contrast to the PEPSS-ASD (Starr, 2000), items 21-28 on

the CSS utilized a 6-point scale to assess parent perceptions of school service provision designed to meet heterogeneous needs (0 for "strongly disagree," 5 for "strongly agree"). Again, Starr's (2000) scale was modified (i.e., a point was added) to force a non-neutral response (Fink, 2009).

Because the PEPSS-ASD (Starr, 2000) was created for use by parents of children in British schools, further modifications were made to several questions in order to align information with American educational statutes. The PESS-ASD (Starr, 2000) asked parents questions (62, 64, 100) about written educational plans for their children with disabilities, and about placement of their children into "regular" or "inclusion" classes by governing British agencies. The CSS addressed placement of students with disabilities and written educational plans as well, but used terms and constructs from American law that govern school practices for children with disabilities. According to the Individuals with Disabilities Education Act, P.L. 108-446, 118 Stat. 2647 (IDEA) Part B, a written Individualized Education Plan (IEP) must have been provided and implemented through the school system for every child identified as having a disability (Chedd et al., 2006). The CSS addressed IDEA's IEP requirement through item10, which asked parents whether or not their children with PWS had written IEPs. Moreover, according to IDEA, school systems have been required to qualify children ages 3-21 with certain special needs as having a disability under one of 13 specific categories (e.g., mental retardation, specific learning disability, autism, orthopedic impairment; Individuals with Disabilities Education Act; IDEA; P.L. 108-446, 118 Stat. 2647). The CSS addressed qualification through item 10.a., which asked parents to identify the category under which their children have been qualified for services under IDEA.

In America, children with disabilities have been able to receive modifications to the standard course of study through a 504 Plan (PL 95-602). Typically, students who have had 504 Plans have not had IEPs, because the modifications allotted for by a 504 Plan would have been subsumed by an Individualized Education Program (IDEA; P.L. 108-446, 118 Stat. 2647). Section 504 of the Rehabilitation Act (PL 95-602), commonly referred to as the Americans with Disabilities Act (ADA), has required state and local government agencies to provide equal opportunities to people with disabilities. Based on Section 504, students with disabilities have been able to receive modifications to the standard course of study that are deemed necessary for assessing the curriculum adequately. (e.g., books on tape, seating that is close to the teacher, testing in a distraction free area). The CSS addressed Section 504 of P.L. 94-142 in question 10.b., which asked if children who did not have an IEP received accommodations through a 504 Plan.

Furthermore, some of the questions on the PEPSS-ASD (Starr, 2000), limited the scope of where education can occur when compared to American school practices (Department for Children, Schools, and Families, 2009). In England children with disabilities have been placed in either "mainstreamed" (i.e., with typically developing peers) or "non-mainstreamed" (i.e., only with children with disabilities) schools (Department for Children, Schools, and Families, 2009). Parents have also been able to pay for private school. Questions 90, 92, and 100 of the PEPSS-ASD (Starr, 2000) reflect this British educational service provision for children with disabilities. However, in the United States, the education of children with disabilities has involved different and more varied settings than the British model (Hess, Molina, & Kozleski, 2006). Therefore, when the CSS asked parents to describe their children's classroom placements (question 11), the CSS offered options that have been offered within American schools (e.g., full time regular classroom, full time regular classroom with a one-on-one aide, part time in a resource room; Hess, Molina, & Kozleski, 2006). Both British and American schools have required written educational plans that are created by a team that includes parents; CSS items 34-38 were modeled after PEPSS-ASD (Starr, 2000) items 42-67, which addressed the issue of quality written educational plans. Hence, the CSS was modeled after the PEPSS-ASD (Starr, 2000) when requesting that parents respond to questions about special education service delivery. However, modifications were made to align the CSS with American educational statutes and practices where they differ from those in Britain.

Overall, the CSS method for assessing participant demographics, parent perceptions of individual child needs for support, and educational support services was inspired by the PEPSS-ASD (Starr, 2000). However, the CSS differed from the PEPSS-ASD (Starr, 2000) on several scales of measurement, with regard to content that is specific to PWS, and where British and American educational services differ (Fink, 2009).

As noted, the FSS (Dunst et al., 1988) also served as a model for the development of the CSS concerning methods for assessing perceived support. In particular, items CSS 39-107 generally followed the format of the FSS (Dunst et al., 1988), with a few modifications. The goal of both the CSS and the FSS (Dunst et al., 1988) was to assess how helpful particular people or groups (i.e., sources of support) were perceived by parents raising a child (Dunst et al., 1988). However, the two surveys differed in the number and specificity of items. The FSS (Dunst et al., 1988) consisted of 20 items (i.e., potential support sources) and used a 5-point helpfulness scale (1 being "not at all helpful," and 5 being "extremely helpful"). The FSS also offered a "not available" option, whereas the section of

the CSS that followed the FSS (i.e., items 39-107) used a slightly modified scale (i.e., a 5 point helpfulness scale with "not needed" and "not available" options) and contained many more items (i.e., 68 items).

The increased number of items on the CSS juxtaposed to the FSS facilitated a more specific accounting of the nature of support that families of children with PWS utilized, found helpful, and found available. As noted, the FSS (Dunst et al., 1988) examined perceptions of social support with ten items (e.g., "parents," "own children," "other parents," co-workers"), and examined all other sources of support with ten more items (e.g., "professional helpers," "school/day-care center," and "professional agencies"). Because this study sought to investigate parent perception of the helpfulness of support by school personnel, community (non-school-affiliated) professionals, social networks (family members, friends, and social acquaintances), and printed informational sources, the CSS included 68 specific sources of support that were intended to capture perceptions across the breadth of each support domain. In addition, supports that have been common to PWS were included in the survey. More specifically, school support items were expanded from 2 items to 16 (e.g., including items such as "exceptional children's teacher," "cafeteria staff," and "school-based speech therapist". Community (non-school-affiliated) professional support items were expanded from 6 items to 34 (e.g., including "dietician," "PWS clinic," "social worker," "case manager," "private psychologist," "neuropsychologist," and "psychiatrist"). Social support was expanded from 11 items to 13 including items such as "church members." In addition, five items were added to address informational support such as literature (e.g., "Books," and "Journals") as well as the Internet, which has been an increasingly viable source of information and support for parents of children with disabilities (Baum, 2004). See Tables 1 and 2 for detailed listings of how individual support items fall into the four categories of sources of support

Another difference between the FSS (Dunst et al., 1988) and the new measure was that the CSS provided "not needed" and "not available" respondent options. The FSS (Dunst et al., 1988) only offered a "not available" option. This modification was thought necessary to differentiate between those who needed a particular support but did not have it, and those who did not need the given support, despite availability.

The FSS (Dunst et al., 1988) did not address barriers to support. However, because barriers to support have typically been present for families with children with PWS (Freedman & Boyer, 2000; Whitman, 2006), an item was added to the CSS (108) for identifying those barriers. CSS item 108 addressed the barriers of financial and informational barriers as well as barriers related to local, qualified providers and uninformed social source of support regarding PWS (Freedman & Boyer, 2000; Whitman, 2006).

With regard to internal consistency reliability, the FSS (Dunst et al., 1988) has been indicated at .77; split-half reliability has been indicated at .75; and test-retest reliability (one month interval) has been indicated at .75 to .91 (Dunst, 1985). Test-retest reliability after 1 to 2 years yielded an average of $r = .50$ for the total score.

Due to the newness of the measure, reliability was not established for the CSS prior to the study. However, analysis included Cronbach's Alpha for the CSS to determine the internal consistency reliability of this instrument with the given PWS sample (reported below).

The Global Assessment of Individual's Behavior–Prader-Willi Syndrome (GAIB-PWS; Tasse et al., 2002) was also used to inform the content of the CSS in regard to PWS child maladaptive behavior symptoms. The GAIB-PWS was designed to assess the behavioral problems of individuals with PWS that interfere with functioning in daily settings, including school settings. Hence, the GAIB-PWS informed CSS questions (13-28) which assessed PWS-related child behavioral concerns and school support regarding those behaviors (e.g., support for following a food plan). In particular, GAIB-PWS items 2-3 and 41-64 addressed the need for children with PWS to have special food plans and exercise, and informed CSS items 13-14 and 21-22. GAIB-PWS items 19, 26, and 62 addressed anxious behaviors and informed CSS items 15 and 23; GAIB-PWS items 16, 23, 28, 52, and 57 addressed moody and sad affect and informed CSS items 16 and 24; and GAIB-PWS items 51, 52, and 57 addressed angry and frustrated behaviors and informed CSS items 17 and 25. In addition, GAIB-PWS items 35-40 addressed atypical behaviors and informed CSS items 18 and 26; GAIB-PWS items 65-80 and A-D addressed obsessive and compulsive behaviors and informed CSS items 19 and 27, and GAIB-PWS items 1, 45, 48, and 61 addressed rule following behaviors and informed CSS items 20 and 28. Taken together, the PEPSS-ASD (Starr, 2000), FSS (Dunst et al., 1988), and GAIB-PWS (Tasse et al, 2002) were the primary sources used to inform the CSS, both in form and content. Table # describes original survey purpose and contribution to the CSS for the noted three contributing surveys. Alterations to original surveys are also reported, as are reliability and validity, where available.

CSS items 72 ("Other School Personnel") and 107 ("Other") were dropped due to missing data > 10%. Helpfulness rating means were computed for each support construct (17 educational support source items, $M = 3.36$; 33 professional support source items, $M = 3.37$; 13 social support source items, $M = 2.92$; 4 informational support source items, $M = 3.30$). Cronbach's Alpha was computed as an estimate of internal reliability on the four means ($n = 71$, $\alpha = .675$; $n = 61$, $\alpha = .641$). The estimation of reliability is lower than preferred, but reasonable due to reliability

being calculated on only 4 items. In addition, "NN" and "Nav" responses were not included in analyses for research questions 2 and 3. Reliability could not be computed on the surveys per item, due to the rating scale including qualitative and quantitative response options.

Appendix J

Categories of Sources of Support and Individual CSS Support Source Items

Educational	Regular education teacher(s), Special education teacher(s), Teacher assistant(s), Special education facilitator, School nurse, One-on-one school aide, School psychologist, School counselor, School administrator, Cafeteria staff, Bus driver, Tutor(s), School-based social worker, dietician, or occupational, speech, or physical therapist, (Other)
Professional	Parent support group, Other support group, Minister/religious leader, Physician, Child's pediatrician, Specialized medical professionals, Child's dentist, Summer camp staff, Early childhood intervention program, Private psychologist, Family therapist, Neuropsychologist, Psychiatrist, Non-school-based social worker, dietician, or occupational, speech, or physical therapist, Case manager, Professional respite care, Financial counselor, Developmental therapist, Department of Social Services, Prader-Willi clinic, Other clinic(s) Foundations/associations for PWS, Foundations/associations for other developmental disabilities, Hotline, Parent workshops/retreats, Layer/legal advocate, Professional parent-to-parent or family-to-family support, Extra curricular activities, (Other)
Social	Parents, Spouse/partner's parents, Relatives/kin, Spouse's relatives/kin, Spouse /partner, Friends, Spouse /partner's friends, Own children, Other parents, Co-workers, Babysitter, Social groups/clubs, Church members, (Other)
Informational	Books on PWS, Medical journals, Internet information on PWS, Pamphlets on PWS, (Other)

Appendix K
Survey Contributions to the CSS, Alterations, and Reliability and Validity

Contributing Survey	Purpose of Survey	Reliability and Validity	Original Survey/ Document's Contribution to CSS	Alterations made to Original Survey
Parent Education Perception and Satisfaction Survey-Autism Spectrum Disorder (PEPSS-ASD; Starr, 2000).	Used in studies to assess the perceptions of parent of children on the autism spectrum with regard to support and services by school personnel (Starr & Foy; 2006; Starr, Foy & Cramer; 2001).	Used in published studies (Starr, & Foy 2006). Reliability and validity information is not currently available.	Demographic, general and special educational, and medical information (PEPSS-ASD; 72-85; 90; 93, 100).	Altered to suit American law and school practices and the PWS population; decreased amount of information requested (CSS; 1-12); omissions of some PEPSS-ASD demographic information (PEPSS-ASD; 1-2, 72-76, 86-87, 89, 94, 96-106).
			Methodology for surveying hetero- and homogeneous symptomology (PEPSS-ASD; 4-33).	Altered symptomology to match that of PWS (CSS; 13-38).
				Scale altered from levels of agreement to "yes" "no" (CSS; 13-24).
				Scale altered from 5 levels of agreement to 6 to avoid a neutral choice (CSS; 24-38; Fink, 2009).
			Parent satisfaction items (PEPSS-ASD; 68).	Expanded parent satisfaction section to include four types of support (CSS; 109-112).
				Scale alteration from a 5- to a 4-point scale to force a non-neutral choice (CSS; 109-112; Fink, 2009).

Family Support Scale (FSS; Dunst, Jenkins, & Trivette, 1988).	Used to assess how helpful particular people or groups (i.e., sources of support) are to a family in raising a young child (Dunst, Jenkins, & Trivette, 1988; Hassal, Rose, & McDonald, 2005).	Internal consistency reliability: .77; split-half reliability: .75; test-retest reliability (1 month interval): .75 to .91 (Dunst, 1985).	Methodology for assessing support received and perception of helpfulness scale (FSS; 1-20).	Increased number of items to more specifically and broadly account for nature and helpfulness of support received; school support was expanded from 2 items to 16; community (non-school-affiliated) professional support from 6 to 34; social support from 11 to 13; and printed/internet information from 0 to 5 (CSS; 39-107).; scale was slightly modified to account for support that is not available.
				Offers "not needed" and "not available" response choices discriminate between needed and not needed unavailable supports (CSS; 39-107).
Global Assessment of Individual's Behavior–Prader-Willi Syndrome (GAIB-PWS; Tasse, Havercamp, Mandal, 2002).	Measures social competence, food and non-food related behavioral challenges, and obsessive and compulsive behaviors for individuals with PWS.	The GAIB-PWS is beginning to be used in studies (Schoch, Powell, Callanan, Havercamp, & Tasse, 2006). Reliability and validity information is currently not available.	Informed items in regard to PWS behavioral symptomology (GAIB-PWS; 1, 2-3, 16, 19, 23, 28, 26, 35-80, A-D).	Content for surveying hetero- and homogeneous symptomology (CSS; 13-28).

Appendix L

Constructs Derived from Parent and Child Measures

Constructs Derived from Parent Measures

Construct	Measure	Items	Scores Utilized	Reliability	Other Support for Use
Parent Measures: Support Constructs (Research Question 1-3; a through d)					
a. Support - school	CSS	61-63, 63a, 64-72, 77, 79, 81, 83, 85 (total = 17)	*M* of raw scores for items that are not NN or Nav	Cronbach's Alpha internal consistency reliability: α = .675	Questions were modified from FSS (FSS; Dunst, Jenkins, & Trivette, 1988) and the PEPSS-ASD, (Starr, 2000) which have been used in published studies.
b. Support - non-school-affiliated professionals	CSS	50-51, 53, 55- 60, 73-76, 78, 80, 82, 84, 86-96, 102-106 (total = 33)	*M* of raw scores for items that are not NN or Nav	See "Support-school."	See "Support-school."
c. Support – social	CSS	39-49, 52, 54 (total = 13)	*M* of raw scores for items that are not NN or Nav	See "Support-school"	See "Support-school"
d. Support – print informational sources	CSS	97, 98, 100, 101 (total = 4)	*M* of raw scores for items that are not NN or Nav	See "Support-school"	See "Support-school"
Parent Measures: Parent Stress					
Parent stress	PSI/SF, SF (Abidin, 1997)	All Items (1-36)	Total Parent Stress Score	Internal reliability .80 to .91; test-retest reliability .68 to .85. Cronbach's Alpha internal consistency reliability: α = .954	Normed on large sample (Abidin, 1997); has been utilized in many studies on parents of children with developmental disabilities (Aunos, Feldman, & Goupil, 2008; Wheeler, Hatton, Reichart, & Bailey, 2007).
Parent Measures: Descriptions of Support Satisfaction and Barriers					
Satisfaction with support	CSS	109-112	Descriptive Statistics	N/A	Questions were modified from the PEPSS-ASD, (Starr, 2000) which has been used in published studies.

Barriers to support	CSS	108	Descriptive Statistics	N/A	Similar barriers have been sighted (Freedman & Boyer, 2000; Whitman, 2006).
Parent Measures: Descriptions of School Services and Family Demographics					
Description of school practices and services	CSS	9-12	Descriptive Statistics	N/A	See "Family and child demographics and general descriptions."
Family/child demographic descriptions	CSS	1-8; 13-20	Descriptive Statistics	N/A	Questions were modified from the PEPSS-ASD, (Starr, 2000) which has been used in published studies.

Constructs Derived from Child Measures

Construct	Measure	Items	Scores Utilized	Reliability	Support for Use
Child Measures: Behavior					
Maladaptive behavior	Global Assessment of Individual's Behavior–Prader-Willi Syndrome (GAIB-PWS, Tasse, Havercamp, Mandal, 2002).	1-80, (#s 1-10 will be reversed scale) and items A-D ("yes=1; "no"=0)	Total raw Score	Cronbach's Alpha provided an estimate of internal consistency reliability (α = .975, n = 61)	Widely used clinically; beginning to be used in studies.
Child Measures: Intelligence					
Intelligence	Stanford Binet- V; routing tests (Roid, 2003)	Items from 2 routing subtests	Abbreviated IQ Score	Reliabilities: .84 to .89 (Roid, 2003).	Normed on large sample; widely utilized; Good estimate of FSIQ

Appendix M

Research Questions, Statistical Procedures, and Exploration to Meet Procedure Assumptions

	Survey Source	Statistical Procedure	Procedure's Use	Key Assumptions of Procedure	Assumptions Explored
Q1	CSS	Frequency Distributions	Frequency of responses; percentages	Descriptive/ exploratory	N/A
Q2	CSS	Simultaneous Repeated Measures ANOVA	Indicated if there was a significant difference between at least two means while reducing the error variance caused by between-group differences	Variance/ covariance matrix of the dependent variables for each "measure" is circular in form [a]	Bartlett's Test of Sphericity; or report test statistics that do not assume sphericity[a]
		(If significant F value) Bonferroni post hoc method	Determined the of the minimum difference between treatment means that is necessary for significance while controlling for Type I error rate	Same sample size across treatments	Completion of surveys and missing data patterns
Q3	CSS, PSI/SF GAIB, and SB-Vr	Multiple Regression	Indicated prediction relationship between independent (predictor) variables and the dependent (criterion) variable	Normality	Examination of histograms and descriptive statistics
				Low multi-collinearity	Examination of Tolerance/ VIFs

a Tabachnik and Fidell (2007, p. 281) note that when using Simultaneous Repeated Measures ANOVA, IVs may be utilized without concern for survey counterbalancing or violation of sphericity.

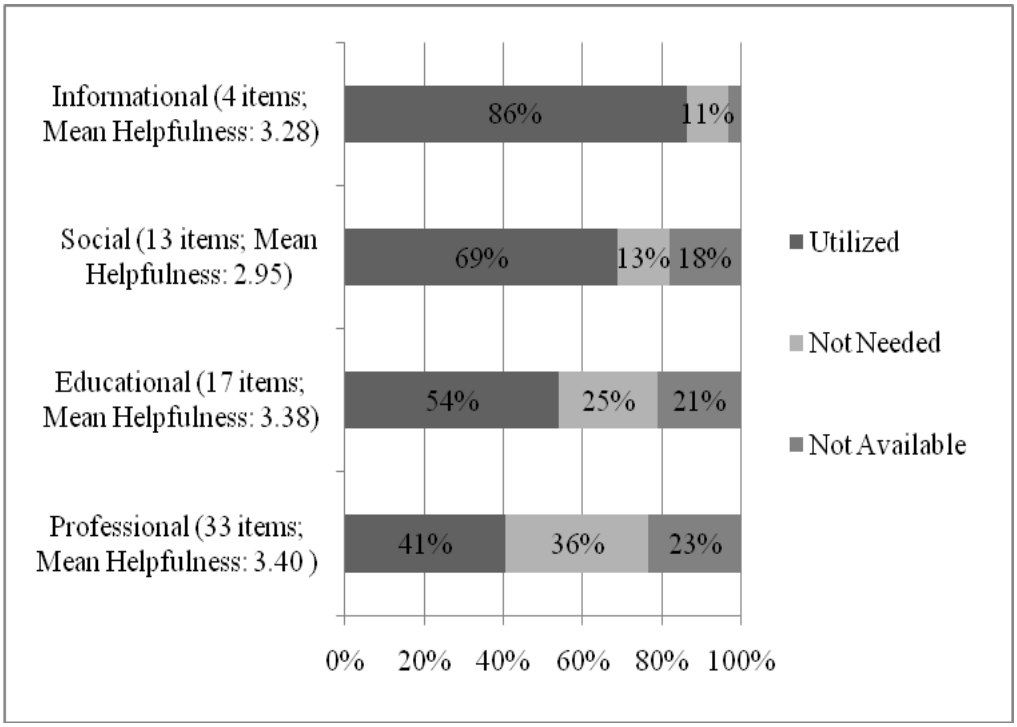
Appendix N
Descriptive Scores for Variables Included in Analyses

Variable	*N*	*Mean*	*SD*	*Range*	*Skewness*	*Kurtosis*
Educational Support Mean	71	3.38	.87	1.00 - 5.00	-.317	-.712
Helpfulness (CSS)	61	3.31	.89	1.30 – 4.75	-.304	-.831
Professional Support Mean	71	3.40	.66	1.06- 4.06	-.457	-.137
Helpfulness (CSS)	61	3.31	.70	1.86 – 4.86	-.326	-.682
Social Support Mean	71	2.95	.71	1.06 – 4.06	.442	-.236
Helpfulness (CSS)	61	2.92	.71	1.63 - 4.60	.359	-.438
Informational Support Mean	71	3.28	.97	1.50 – 5.00	.213	-.652
Helpfulness (CSS)	61	3.28	.99	1.50 - 5.00	.163	-.716
Parent Stress (PSI total stress)	61	89.34	26.67	36 - 161	.374	.309
Child Maladaptive Behavior	61	123.52	68.01	34 - 300	.732	-.061
(GAIB-PWS total score)						
Child Age (years)	61	10.21	4.23	3 – 19[a]	.289	-.943
Abbreviated IQ	26	84.27[b]	19.25	47 – 121[b]	-.319	-.650
(SB-Vr standard score)	21	78.67[c]	16.74	47 – 100[c]	-.439	.972

[a] Child age range at time of administration of SB-Vr: 3 – 16 years.
[b] Abbreviated IQ of 121 is an outlier in comparison to PWS population where the majority of individuals score between 50-70 (Butler et al., 2006; Kundert, 2008; Whittington et al., 2004; Whitman & Thompson, 2006).
[c] Abbreviated IQ descriptive without 5 cases (n = 21) where scores feel above 100 (103x2, 106 x2, 121x1).

Appendix O
Research Question 1 Results

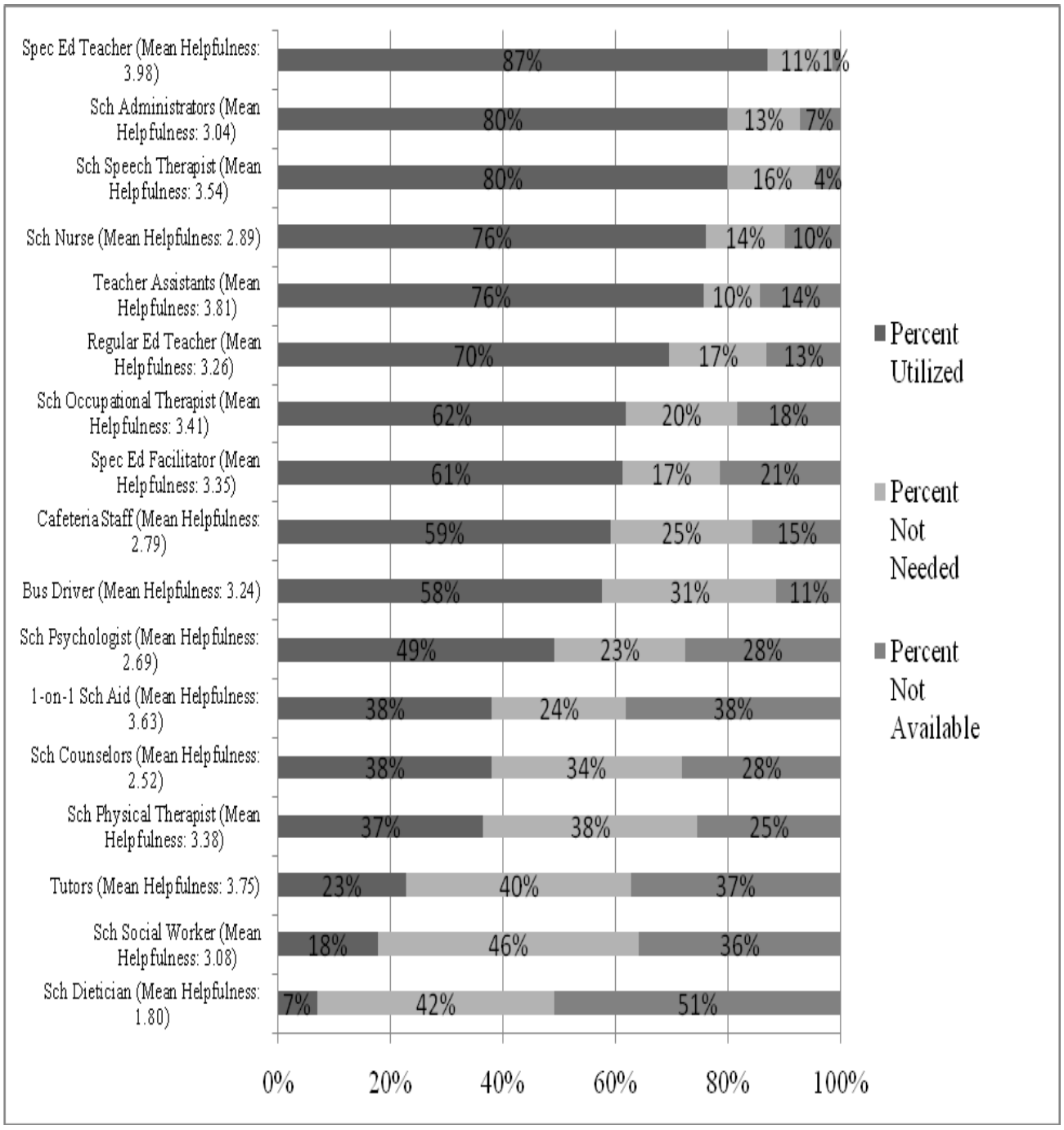
Support Source Category Percentages for Supports Perceived as Utilized, Not Needed, and Not Available; and Mean Helpfulness Ratings for Sources Rated as Utilized[a]



[a]Mean helpfulness data were calculated using only support sources that were rated on the CSS helpfulness scale (scores of 1 through 5); "Nav" and "NN" responses were not on the helpfulness scale and therefore were not used in mean helpfulness calculations.

Appendix O
Research Question 1 Results

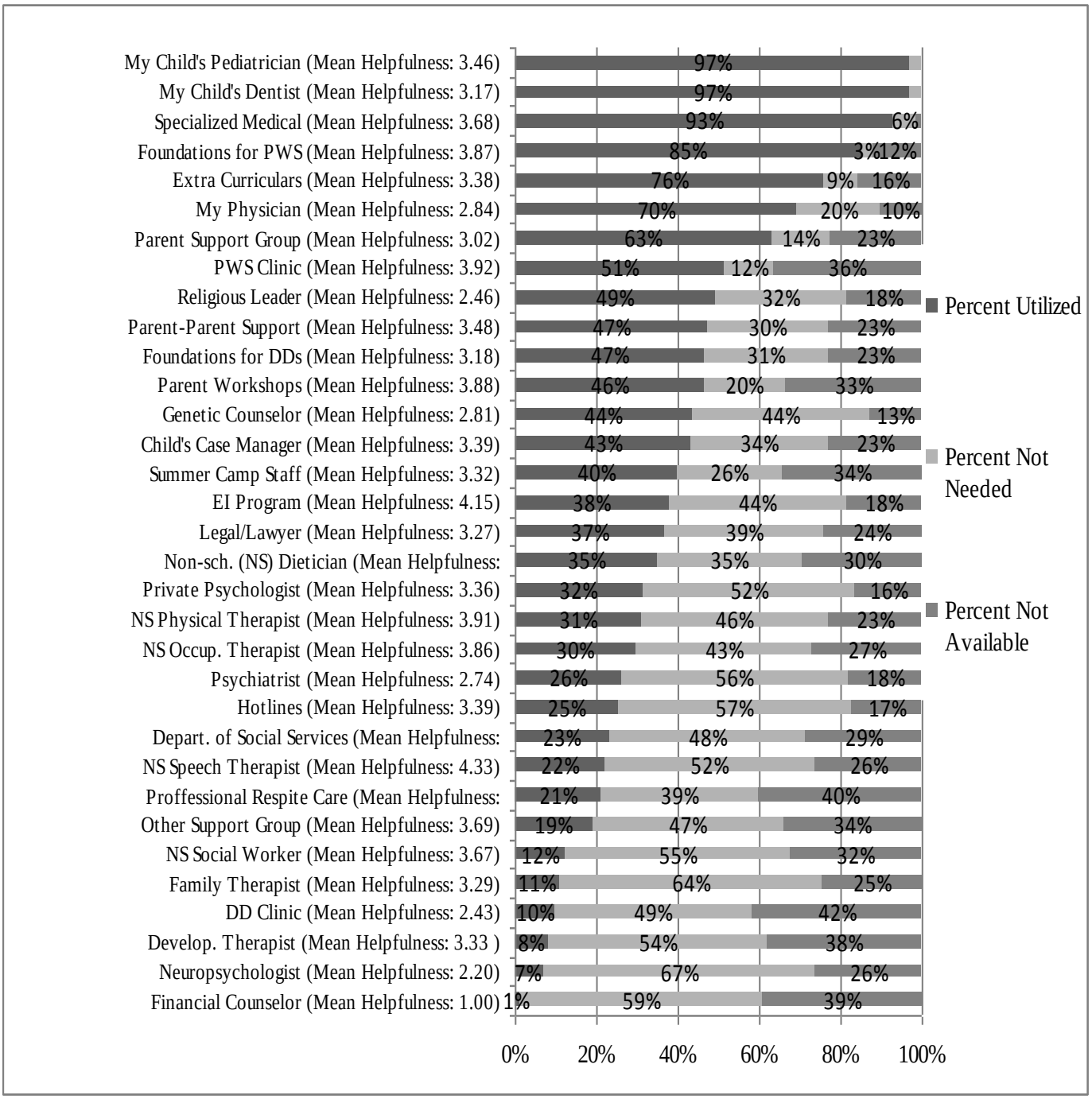
Educational Support Source Percentages for Supports Perceived as Available and Utilized, Not Needed, and Not Available; and Mean Helpfulness Ratings for Sources Rated as Utilized[a]



[a]Mean helpfulness data were calculated using only support sources that were rated on the CSS helpfulness scale (scores of 1 through 5); "Nav" and "NN" responses were not on the helpfulness scale and therefore were not used in mean helpfulness calculations.

Appendix O
Research Question 1 Results

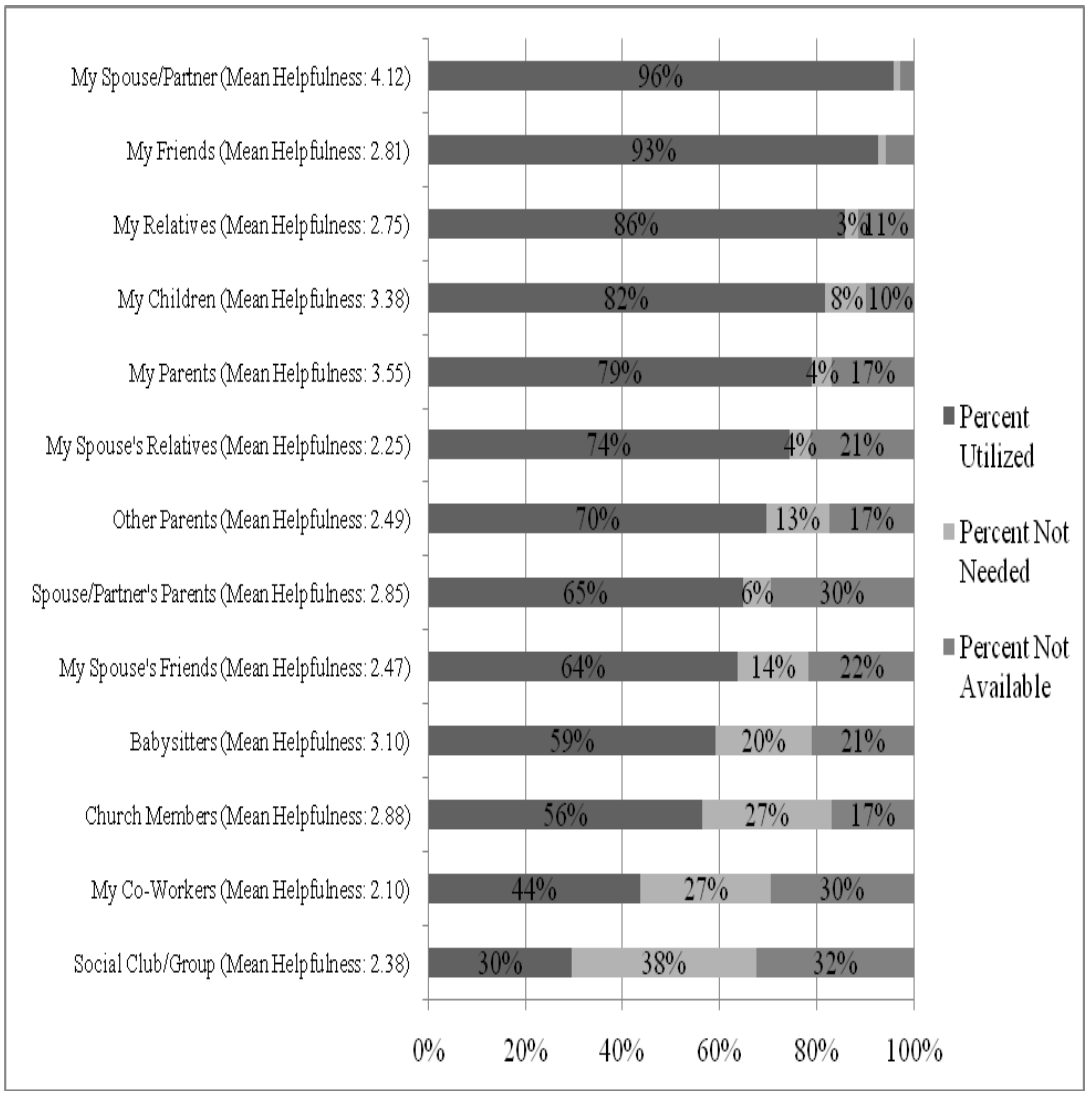
Professional Support Source Percentages for Supports Perceived as Available and Utilized, Not Needed, and Not Available; and Mean Helpfulness Ratings for Sources Rated as Utilized[a]



[a]Mean helpfulness data were calculated using only support sources that were rated on the CSS helpfulness scale (scores of 1 through 5); "Nav" and "NN" responses were not on the helpfulness scale and therefore were not used in mean helpfulness calculations.

Appendix O
Research Question 1 Results

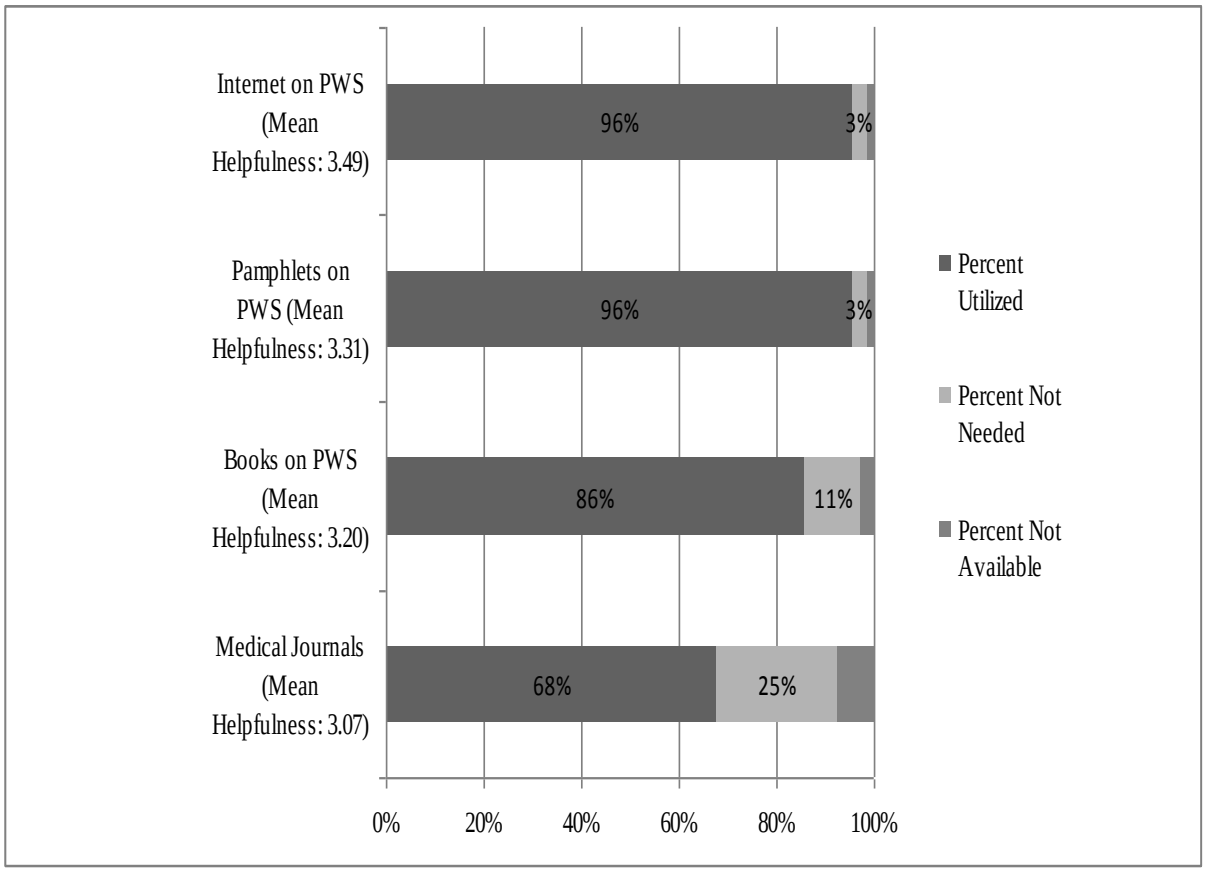
Social Support Source Percentages for Supports Perceived as Available and Utilized, Not Needed, and Not Available; and Mean Helpfulness Ratings for Sources Rated as Utilized[a]



[a]Mean helpfulness data were calculated using only support sources that were rated on the CSS helpfulness scale (scores of 1 through 5); "Nav" and "NN" responses were not on the helpfulness scale and therefore were not used in mean helpfulness calculations.

Appendix O
Research Question 1 Results

Informational Support Source Percentages for Supports Perceived as Available and Utilized, Not Needed, and Not Available; and Mean Helpfulness Ratings for Sources Rated as Utilized[a]



[a]Mean helpfulness data were calculated using only support sources that were rated on the CSS helpfulness scale (scores of 1 through 5); "Nav" and "NN" responses were not on the helpfulness scale and therefore were not used in mean helpfulness calculations.

Appendix O
Research Question 1 Results

Mean Perceived Helpfulness per Individual Support Sources and Number Who Utilized Sources (n = 71)

Category of Support	Support Source Item	Mean Rating from 1-5	Number Who Utilized This Support
Educational	Special education teacher(s)	3.98	61
	Teacher assistant(s)	3.81	53
	Tutor(s)[a]	3.75	16
	One-on-one school aide[a]	3.63	27
	School-based speech therapist	3.54	56
	School-based occupational therapist	3.41	44
	School-based physical therapist[a]	3.38	26
	Special education facilitator	3.35	43
	Regular education teacher(s)	3.26	48
	Bus driver	3.24	41
	School-based social worker[a]	3.08	12
	School administrator	3.04	56
	School nurse	2.89	54
	Cafeteria staff	2.79	42
	School psychologist	2.69	34
	School counselor[a]	2.52	27
	School-based dietician[a]	1.80	5
Professional	Non-school-based speech therapist[a]	4.33	16
	Early childhood intervention program[a]	4.15	27
	Prader-Willi clinic	3.92	38
	Non-school-based physical therapist[a]	3.91	23
	Parent workshops/retreats	3.88	32
	Foundations/associations for PWS	3.87	64
	Non-school-based occupational therapist[a]	3.86	22
	Professional respite care[a]	3.80	16
	Other support group[a]	3.69	13
	Specialized medical professionals	3.68	67
	Non-school-based social worker[a]	3.67	9
	Professional parent-to-parent or family-to-family support	3.48	33
	Child's pediatrician	3.46	68
	Case manager	3.39	32
	Hotline[a]	3.39	16
	Extracurricular activities	3.38	53
	Private psychologist[a]	3.36	23
	Developmental therapist[a]	3.33	6
	Summer camp staff	3.32	28
	Family therapist[a]	3.29	8
	Layer/legal advocate[a]	3.27	26
	Foundations for those w/ developmental disabilities	3.18	35
	Child's dentist	3.17	68
	Non-school-based dietician[a]	3.16	25
	Parent support group	3.02	45
	My Physician	2.84	48
	Genetic Counselor	2.81	31
	Psychiatrist[a]	2.74	19

	Department of Social Services[a]	2.71	17
	Minister/religious leader	2.46	35
	Clinics for those with DDs[a]	2.43	7
	Neuropsychologist[a]	2.20	5
	Financial counselor[a]	1.00	1
Social	Spouse /partner	4.12	67
	My parents	3.55	56
	Own children	3.38	58
	Babysitter	3.10	42
	Church members	2.88	40
	Spouse/partner's parents	2.85	46
	Friends	2.81	63
	My relatives/kin	2.75	60
	Other parents	2.49	48
	Spouse /partner's friends	2.47	44
	Social groups/clubs[a]	2.38	21
	Spouse's relatives/kin	2.25	52
	Co-workers	2.10	31
Informational	Internet information on PWS	3.49	66
	Pamphlets on PWS	3.31	66
	Books on PWS	3.20	60
	Medical journals	3.07	46

[a] Less than 40% of participants indicated using these support sources (i.e., they were rated "NN" or "Nav" 60% or more).

Appendix O
Research Question 1 Results

Parent Reported Barriers to Support (n=71)

Reported Barrier	*n*	*%*
Lack of Adequate Knowledge: Mental Health Professionals	37	55.2%
Lack of Agencies w/ Appropriate Resources	32	47.8%
Lack of Time	31	46.3%
Lack of Adequate Knowledge: School Personnel	29	43.3%
Lack of Money	28	41.8%
Lack of Adequate Knowledge: Medical Professionals	28	41.8%
Lack of Adequate Childcare	18	26.9%
Feelings That Family Should Handle Issues w/out Outside Help	13	19.4%
[a]Other	13	19.4%
Lack of Adequate Health Insurance	6	8.6%
(Perceived) Prejudice Towards Family due to PWS	5	7.5%
Unsure Where Find Information	4	6.0%
Lack of Adequate Transportation	3	4.8%
Lack of Computer/Internet Access	1	1.5%
My Own Disability	1	1.5%
Language Barrier	0	0.0%

[a]Written in responses included: "Make too much money for Medicaid," "I have 4 children," "Lack of self-knowledge; I should know more," "My family feels my son should be my responsibility and that I should handle him on my own," "Not enough community activities," "[No] families with a child with PWS in close proximity," and "[We've been turned down] for CAP [Client Assistance Program] services."

Reported Satisfaction with Support: Percentage by Domain (n = 71)

Support Category	Completely Satisfied	Fairly Satisfied	Mildly Satisfied	Not Satisfied
Educational Personnel	36.4%	40.9%	12.1%	10.6%
Professionals Outside the School Setting	17.7%	61.8%	14.7%	5.9%
Family, Friends, and Social Network	26.5%	47.1%	16.18%	10.3%
Printed Information (including internet sources)	17.7%	54.4%	25.0%	2.9%

Appendix P
Research Question 2 Results

Mean Helpfulness per Category for Support Sources That Were Utilized by 40% or More Participants (n = 65)

Support Category[a]	*Mean*	*SD*
Educational Support	3.36	.899
Professional Support	3.33	.707
Social Support	2.94	.727
Informational Support	3.31	.985

[a]Respondents rated these support sources with a combined percentage of greater than 60% NN and Nav. For these support sources, 29 or fewer participants utilized those supports within the 6 months prior to completing the survey. Because so few respondents utilized those supports, these individual support services were dropped from question two and three analyses, which utilized the variables of mean helpfulness of utilized supports across categories.

Analysis of Variance Summary (N = 65)

Correction	Sum of Squares	df	Mean Square	F	Significance	Partial Eta Squared
Huynh-Feldt[a]	7.666	2.263	3.388	5.654**	.003	.081[b]
Error	86.782	144.828	.599			

**p < 0.01

[a]Tabachnick and Fidell (2007, p. 281) note that violation of sphericity is not a consideration with simultaneous repeated measures ANOVAs. However, a correction for violation of sphericity (i.e., Huynh-Feldt) was used for a conservative estimate of omnibus effects.

[b] η^2_L was used as a lower bound of effect size due to violation with sphericity; effect size was identical for Repeated Measures ANOVA statistic ran with sphericity assumed.

Bonferroni Comparison for Helpfulness of Support per Category (N = 65)

Support Type	Support Type	Mean Difference	Std. Error	95% Confidence Interval for Difference	
				Lower Bound	Upper Bound
Educational	Professional	.024	.086	-.211	.259
	Social	.419*	.117	.101	.737
	Informational	.049	.135	-.318	.415
Professional	Educational	-.024	.086	-.259	.211
	Social	.395*	.096	.133	.656
	Informational	.025	.103	-.256	.305

* p < 0.05

Appendix Q
Research Question 3 Results

Pearson Correlation Matrix of Parent Perceived Stress and Support, and Child Behavior and Age (N = 61)

			Perceived Helpfulness of Support Categories (Utilized Supports)					
	Parent Stress	Child Mal. Behavior	Educational	Professional	Social	Informational	Child Age	Child Abbrev. IQ
Parent Stress		.669**	-.283*	-.310*	-.374**	-.194	.194	-.133
Child Behavior			-.130	-.135	-.427**	.010	.176	-.117
Educational				.621**	.293*	.296*	-.135	-.075
Professional					.370**	.530**	-.084	-.285
Social						-.141	-.136	.047
Informational							-.105	-.158
Child Age								-.487*
Child. IQ								

**p < 0.01. *p < 0.05.

ANOVA for the Regression Equation, Parent Stress on Child Behavior, and Age, and Perceived Helpfulness of Educational, Professional, Social, and Informational Support (N= 61)

	Sum of Squares	df	Mean Square	F	Significance
Regression	22143.881	6	3690.647	9.701**	.000
Residual	20543.890	54	380.442		
Total	42687.770	60			

**p < 0.01.

Summary of Regression Analysis for Parent Stress on Child Behavior and Age, and Perceived Helpfulness of Educational, Professional, Social, and Informational Support (N = 61)

Variable	*B*	*SE B*	*β*	Significance
Child Maladaptive Behavior	.239	.041	.609**	.000
Educational Support Helpfulness	-2.883	3.657	-.096	.434
Professional Support Helpfulness	-2.195	5.783	-.057	.706
Social Support Helpfulness	-2.966	4.637	-.079	.525
Informational Support Helpfulness	-3.986	3.361	-.148	.241
Child Age	.271	.615	.043	.662
R^2		.519		
F for change in R^2		9.701		

*p < .05. **p < .01.

Appendix Q
Research Question 3 Results

ANOVA for the Regression Equation, Parent Stress on Child Behavior, Age, and IQ, and Perceived Helpfulness of Educational, Professional, Social, and Informational Support (N = 26)

	Sum of Squares	df	Mean Square	F	Significance
Regression	14236.346	7	2033.764	3.693**	.012
Residual	9911.808	18	550.656		
Total	24148.154	25			

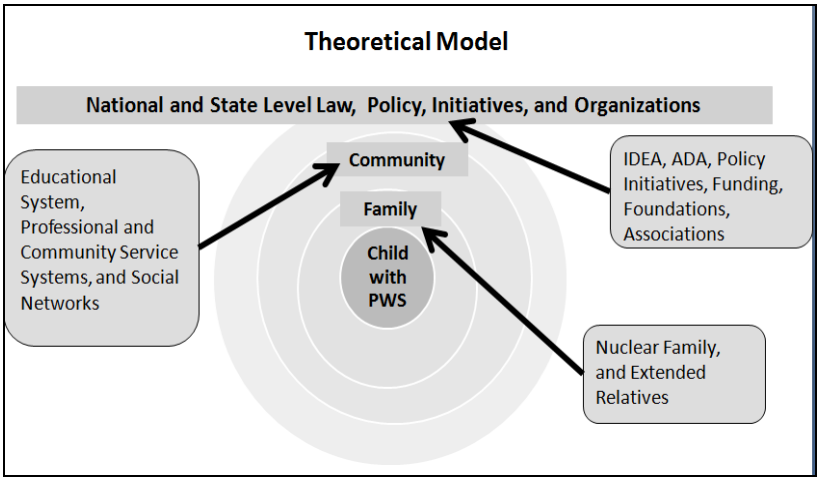
**p < 0.01.

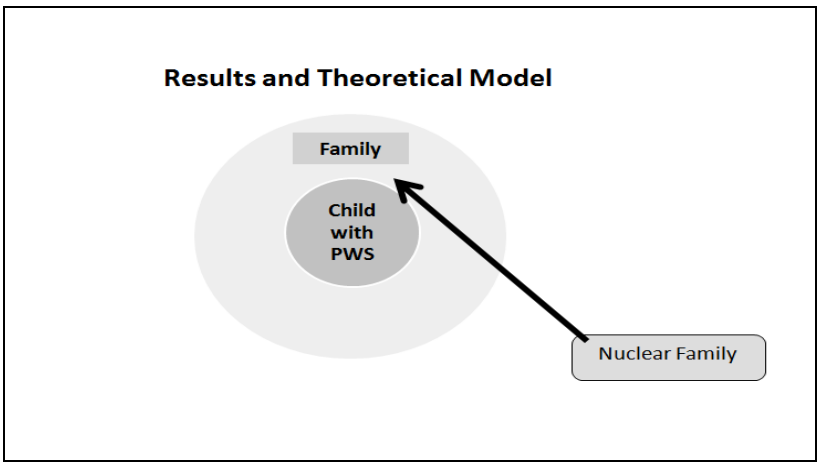
Summary of Regression Analysis for Parent Stress on Child Behavior, Age, and IQ, and Perceived Helpfulness of Educational, Professional, Social, and Informational Support (N = 61)

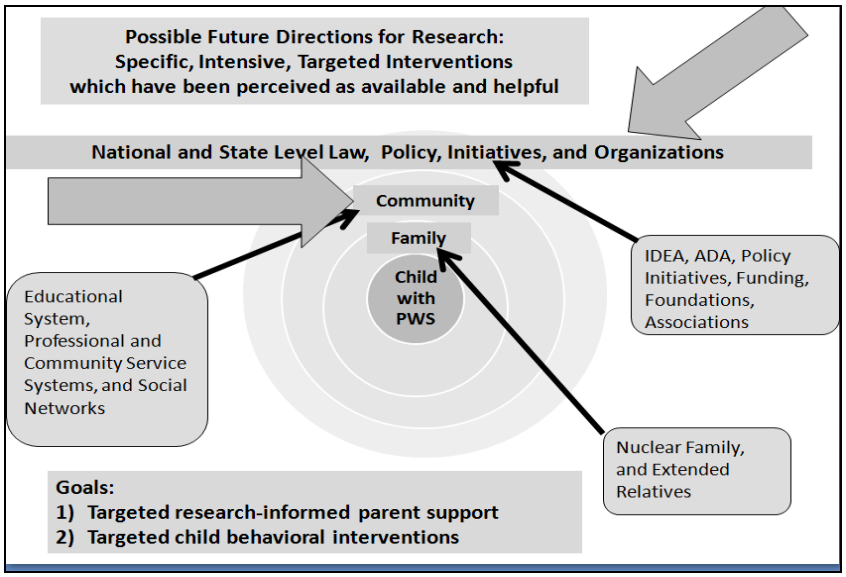
Variable	B	SE B	β	Significance
Child Maladaptive Behavior	.317	.106	.697**	.008
Educational Support Helpfulness	-8.109	5.976	-.252	.192
Professional Support Helpfulness	6.238	12.306	.124	.618
Social Support Helpfulness	2.835	10.278	.063	.768
Informational Support Helpfulness	3.647	6.858	.113	.601
Child Age	1.752	1.500	.228	.258
Child Abbreviated IQ	.149	.312	.092	.640
R2		.590		
F for change in R2		3.693		

*p < .05. **p < .01.

Appendix R
Theoretical Model, Results, and Implications for the Future







References

Abidin, R. R. (1995). Parenting stress index, (3rd ed.): Professional manual. Odessa: Psychological Assessment Resources, Inc.

Abidin, R. R. (1997). Parenting stress index: a measure of the parent-child system. In Zalaquett, C.P. & Wood, R. (Eds.), *Evaluating stress: A book of resources* Lanham: Scarecrow Press, Inc.

Aman, M.G., Tassé, M.J.,Rojahn, J. & Hammer, D. (1996). The Nisonger CBRF: A child behavior rating form for children with developmental disabilities. *Research in Developmental Disabilities, 17*, 41-57.

Atkin K., & Lorch, M. P. (2007). Language development in a 3-year-old boy with Prader-Willi syndrome. *Clinical Linguistics and Phonetics, 21*(4), 261-276.

Aunos, M., Feldman, M., & Goupil, G. (2008). Mothering with intellectual disabilities: relationship between social support, health and well-Being, parenting and child behavior outcomes. *Journal of Applied Research in Intellectual Disabilities, 21,* 320-330.

Baron, R. M., & Kenny, D. A. (1986). The moderator-mediator variable distinction in social psychological research: conceptual, strategic, and statistical considerations. *Journal of Personality and Social Psychology, 51*(6), 1173-1182.

Baum, L. S. (2004). Internet parent support groups for primary caregivers of a child with special health care needs. *Pediatric Nursing, 30*(5), 381-401.

Beck, A., Hastings, R. P., Daley, D. (2004) Pro-social behavior and behavior problems independently predict maternal stress. *Journal of Intellectual and Developmental Disability, 29*(4), 339-349.

Becker, K. A. (2003). *History of the Stanford-Binet intelligence scales: content and psychometrics.* (Stanford-Binet Intelligence Scales, Fifth Edition Assessment Service Bulletin No. 1). Itasca, IL: Riverside Publishing.

Berument, S. K., Rutter, M., Lord, C. Pickles, A., & Bailey, A. (1999). Autism screening questionnaire: diagnostic validity, *The British Journal of Psychiatry, 175,* 444-451.

Boyd, B. A. (2002). Examining the relationship between stress and lack of social support in mothers with children with autism. *Focus on Autism and Other Developmental Disabilities, 17*(4), 208-215.

Bronfenbrenner, U. (1979). *The Ecology of Human Development: Experiments by Nature*

and Design. Cambridge: Harvard University Press.

Buiting & Horsthemke, (2006). Molecular genetic findings in Prader-Willi syndrome. In Butler, M. G., Lee, P. D. K., Whitmen, B. Y. (Eds.), *Management of Prader-Willi Syndrome* (3rd ed., pp. 58-73). New York: Springer.

Butler, M. G., Hanchett, J. M., & Thompson, T. (2006). Clinical Findings and Natural History of Prader-Willi syndrome. In Butler, M. G., Hanchett, J. M., & Thompson, T. (Eds.), *Management of Prader-Willi Syndrome.* (3rd ed., pp. 3-48). New York: Springer.

Cassidy, S. B., & Driscoll, D. J. (2009). Prader-Willi syndrome. *European Journal of Human Genetics 17*, 3-13.

Chedd, N., Levine, K., & Wharton, R. H., 2006. Educational Considerations for Children with Prader-Willi Syndrome. In Butler, M. G., Hanchett, J. M., & Thompson, T. (Eds.), *Management of Prader-Willi Syndrome.* (3rd ed., pp. 302-316). New York: Springer.

Cook, R. S., Rule, S., Maringer, H. (2003). Parent's evaluation of the usability of a website on recommended practices. *Topics in Early Childhood Special Education, 23*(1) 19-27.

Coolican, J., Bryson, S. E., and Zwaigenbaum, L. (2008). Brief report: data on the Stanford Binet Intelligence Scales (5th ed.) in children with Autism Spectrum Disorder. *Journal of Autism and Developmental Disorders, 38,* 190-197.

Conners, F. A., Rosenquist, C. J., Atwell, J. A., Klinger, L. G. (2000). Cognitive strengths and weaknesses associated with Prader-Willi syndrome. *Education and Training in Mental Retardation and Developmental Disabilities, 35*(4), 441-448.

Crinò, A., Schiaffini, R., Ciampalini, P., Spera, S., Beccaria, L, Benzi, F., et al. (2003). Hypogonadism and pubertal development in Prader-Willi syndrome. *European Journal of Pediatrics, 162,* 327-333.

Davies, B. & Ellison, L. (1995). Improving the quality of schools-ask the clients*? School Organization, 15*(1), 5-12.

Deater-Deckard, K. (2004). *Parenting stress*. New Haven: Yale University Press.

Department for Children, Schools, and Families (2009). *Special education needs (SEN): a guide for parents and carers; revised 2009*. Nottingham: Crown.

DiStephano, C., Dombrowski, S. C. (2006). Investigating the theoretical structure of the Stanford-Binet-Fifth Edition. *Journal of Psychoeducational Assessment, 24*(2), 123-136.

Douma, J., C., H., Dekker, M. C., Koot, H. M., (2006). Supporting parents of youths with intellectual disabilities and psychopathology. *Journal of Intellectual Disability Research*, 50(8), 570-581.

Dunst, C. J., Trivette, C. M, & Jenkins, V. (1984). *Family Support Scale: Reliability and Validity*. Ashville: Winterberry Press.

Dunst, C. J., Trivette, C. M, & Jenkins, V. (1988). Family support scale. In C. J. Dunst, C. M. Trivette and A.G. Deal., *Enabling and Empowering Families*: *Guidelines & Principles for Practice*. Cambridge: Brookline Books.

Dykens, E. M. (2002). Are jigsaw puzzles "spared" in persons with Prader-Willi syndrome? *Journal of Child Psychology and Psychiatry, 43*(3), 343-352.

Dykens, E. M., & Kasari, C. (1997). Maladaptive behavior in children with Prader-Willi syndrome, Down syndrome, and nonspecific mental retardation. *American Journal on Mental Retardation, 120,* 228-237.

Fink, A. (2009). *How to conduct surveys; a step-by-step guide, 4th ed.* Thousand Oaks: Sage Publications, Inc.

Freedman, R. I., Boyer, C. N. (2000). The power to choose: supports for families caring for individuals with developmental disabilities. *Health and Social Work*, 25(1), 59-68.

Gedye, A. (1992). Recognizing obsessive-compulsive disorder in clients with developmental disabilities. *Habilitative Mental Healthcare Newsletter 11,* 73-77.

Goff, B. J. (2006a). Education and social issues for adolescents with Prader-Willi syndrome. In Butler, M. G., Hanchett, J. M., & Thompson, T. (Eds.), *Management of Prader-Willi Syndrome.* (3rd ed., 344-355). New York: Springer.

Goff, B. J. (2006b). Advocacy issue: school discipline and expulsion. In Butler, M. G., Hanchett, J. M., & Thompson, T. (Eds.), *Management of Prader-Willi Syndrome.* (3rd ed., 448-456). New York: Springer.

Green, B. L., Furrer, C., McAllister, C. (2007). How do relationships support parenting? Effects of attachment style and social support on parenting behavior in an at-risk populations. *American Journal of Community Psychology, 40,* 98-108.

Greenswag, L. R., & Alexander, R. C. (2006). Introduction. In Butler, M. G., Hanchett, J. M., & Thompson, T. (Eds.), *Management of Prader-Willi Syndrome.* (3rd ed., xxi-xxii). New York: Springer.

Greeves, N., Prince, E. Evans, D. W. & Charman, T. (2006). Repetitive and ritualistic behavior in children with Prader-Willi syndrome and children with Autism. *Journal of Intellectual Disability Research, 50*(2), 92-100.

Gupta, V. B. (2007). Comparison of parenting stress in different developmental disabilities. *Journal of Developmental and Physical Disabilities, 19,* 417-425.

Guralick, M. J., Hammond, M. A., Neville, B. & Connor, R. T. (2008). The relationship between sources and functions of social support and dimensions of child- and parent-related stress. *Journal of Intellectual Disability Research, 52*(12), 1138-1154.

Hassall, R., Rose, J., & McDonald, J. (2005). Parenting stress in mothers of children with an intellectual disability: the effects of parental cognition in relation to child characteristics and family support. *Journal of Intellectual Disability Research, 49*(6), 405-418.

Hastings, R. P. (2002). Parental stress and behavior problems of children with developmental disability. *Journal of Intellectual and Developmental Disabilities, 27*(3), 149-160.

Haveman, M. van Berkum, G., Reijnders, R., Heller, T. (1997). Difference in service needs, time demands, and caregiving burden among parents of persons with mental retardation across the life cycle. *Family Relations, 46,* 417-425.

Heinemann, J. (2008). Are you a researcher or student who is interested in doing a study on PWS? *The Prader-Willi Syndrome Association-USA.* Retrieved October 15, 2008 from http://www.pwsausa.org/research/StudentTopics.htm.

Hess, R. S., Molina, A. M., Kozleski, E. B., (2006). Until somebody hears me: parent voice and advocacy in special education decision making. *British Journal of Special Education, 33*(3), 148-157.

Hill, C. & Rose, J. (2009) Parenting stress in mothers of adults with an intellectual disability: parental cognitions in relation to child characteristics and family support. *Journal of Intellectual Disability Research (53),*12, 969-980.

Hiraiwai, R., Maegaki, Y., Oka, A., Ohno, K. (2007). Behavioral and psychiatric disorders in Prader-Willi syndrome: a population study in Japan. *Brain and Development 29,* 535-542.

Hodapp, R. M., Dikens, E. M., Masino, L. L., (1997). Families of children with Prader-Willi syndrome: stress-support and relations to child characteristics. *Journal of Autism and Developmental Disabilities, 27*(1), 11-24.

Hodapp, R. M., Fidler, D. J., & Smith A. C. (1998). Stress and coping in families

of children with Smith-Magenis syndrome. *Journal of Intellectual Disability Research, 42*(5), 331-340.

Horton, T. V., & Wallander, J. L. (2001). Hope and social support as resilience factors against psychological distress if mothers who care for children with chronic physical conditions. *Rehabilitation Psychology, 46*(4) 382.

Howell, D. C., (2002). *Statistical methods for psychology* (5th ed.). Pacific Grove: Wadsworth Group.

Individuals with Disabilities Education Act. (2004). P.L. No. 108-446, 118 Stat. 2647.

James, T. N., & Brown, R. L. (1992). *Prader-Willi Syndrome: Home School and Community.* Chapman and Hall: New York.

James, T. N., & Brown, R. L. (1993). Prader-Willi syndrome: quality of life issues in home, school, and community. *Australia and New Zealand Journal of Developmental Disabilities, 18*(4), 253-260.

Jauregi, J., Arias, C., Vegas, O., Alén, F., Martinez, S., et al. (2007). A neuropsychological assessment of frontal cognitive functions in Prader-Willi syndrome. *Journal of Intellectual Disability Research, 51* (5), 350-365.

Johnston, C., Hessl, D., Blasey, C., Eliez, S., Erba, H., et al., (2003). Factors associated with parenting stress in mothers of children with Fragile X syndrome. *Developmental and Behavioral Pediatrics*, *24*(4), 267-275.

Kaufman, A. S., (1994) *Intelligence testing with the WISC-III.* New York: John Wiley and Sons, Inc.

Kenny, D. A. (2008). Mediation and moderation. *David A. Kenny's Homepage*. Retrieved on December 29, 2008 from http://davidakenny.net/cm/mediate.htm.

Kersh, J., Hedvat, T. T., Hauser-Cram, P., & Warfield, M. E., (2006) The contribution of marital quality to the well-being of parents of children with developmental disabilities. *Journal of Intellectual Disability Research, 50*(12), 883-893.

Koenig, K., Klin, A., & Schulz, R. (2004). Deficits in social attribution in Prader-Willi syndrome. *Journal of Autism and Developmental Disorders, 34*(5) 573-582.

Kundert, D. K. (2008). Prader-Willi syndrome. *School Psychology Quarterly, 23*(2), 246-257.

Langkamp, D. L., Lehman, A., & Lemeshow, S. (in press). Techniques for handling missing data in secondary analyses of large surveys. *Academic Pediatrics.*

Lloyd, T., & Hastings, R. P. (2008), Psychological variables as correlates of adjustment in mothers of children with intellectual disabilities: cross-sectional and longitudinal relationships. *Journal Intellectual Disability Research*, *52*(1), 37-48.

Lord, C., Rutter, M., & LeCouteur, A. (1994) Autism diagnostic interview- revised: a revised version of a diagnostic interview for caretakers of individuals with possible pervasive developmental disorders. *Journal of Autism and Developmental Disorders*, *24,* 659-685.

Lord, C., Risi, S., Lamberecht, L. Cook, E. H., Leventhal, B. et al. (2000). Autism Diagnostic Observation Schedule – Generic: a standard measure of social and communication deficits with the spectrum of Autism. *Journal of Autism and Developmental Disorders, 30,* 206-223.

McCandless, S. E., & Cassidy, S. B. (2006). Diagnostic criteria for Prader-Willi syndrome. In Butler, M. G., Hanchett, J. M., & Thompson, T. (Eds.), *Management of Prader-Willi Syndrome.* (3rd ed., pp. 49-57). New York: Springer.

McLeskey, J. Henry, D. (1999) Inclusion: what progress is being made across states? *Teaching Exceptional Children 31(*5) 56-62.

McLesky, J., Tyler, N. C., & Saunders, S. S. (2004) The supply of and demand for special education teachers: a review of research regarding the chronic shortage of special education teachers. *The Journal of Special Education (38)*1, 5-21.

Miller, J. L., Couch, J. A., Schmalfuss, I., He, G., Liu, Y., Driscoll, D. J. (2007), Intracranial abnormalities detected by three-dimensional magnetic resonance imaging in Prader-Willi syndrome. *American Journal* of *Medical Genetics Part A 143*(5), 476-483.

Milner, K. M., Craig, E. E., Thompson, R. J., Veltman, M. W. M., Thomas, N. S., Roberts, S., et al. (2005). Prader-Willi syndrome: intellectual abilities and behavioral features by genetic subtype. *Journal of Child Psychology and Psychiatry, 46*(10), 1089-1096.

Mitchell, D. B., Hauser-Cram, P. (2008). The well-being of mothers of adolescents with developmental disabilities in relation to medical care utilization and satisfaction with healthcare. *Research in Developmental Disabilities, 29,* 97-112.

Moos, R.H., Moos, B.S. (1986), *Family Environment Scale Manual,* 2nd. Palo Alto: Consulting Psychologists Press.

Nachshen, J. S. & Minnes, P. (2005). Empowerment in parents of school-aged

children with and without developmental disabilities. *Journal of Intellectual Disability Research*, *49*(12), 889-904.

Neece, C., & Baker, B. (2008). Predicting maternal parenting stress in middle childhood: to roles of child intellectual status, behavior problems, and social skills. *Journal of Intellectual Disability Research, 52*(12), 1114-1128.

Ogletree, B. T., Fischer, M. A., & Shultz, (1999). Bridging the family professional gap: Facilitating interdisciplinary services for children with disabilities. Springfield: Charles C Thomas Publisher, LTD.

Oliver, C., Woodcock, K. A., & Humphreys, G. W. (2009) The relationship between components of the behavioral phenotype of Prader-Willi Syndrome. *Journal of Applied Research in Intellectual Disabilities (22),* 403-407.

Olstad, R., Sexton, H., Søgaard (2001). The Finnmark study. A prospective population study of the social support buffer hypothesis, specifics stressors and mental distress. *Social Psychiatry and Psychiatric Epidemiology*, *36,* 582-589.

Plant, K. M., & Sanders, M. R. (2007) Predictors of care-giver stress in families of preschool-aged children with developmental disabilities. *Journal of Intellectual Disability Research, 51*(2), 109-124.

Plesa-Skwerer, D., Sullivan, K., Joffre, K., Tager-Flusberg, H (2004). Self concept in people with Williams syndrome and Prader–Willi syndrome. *Research in Developmental Disabilities* 25 119–138

Rassler, S., Rubin, D. B., & Schenker, N. (2008). Incomplete data: diagnosis, imputation, and estimation. In de Leeuw, E. D., Hox, J. J., Dillman, D. A. (Eds.), *International Handbook of Survey Methodology* . (pp. 370-386). New York: Laurence Erlbaum Associates.

Raven, J. C. (1936). *Mental tests used in genetic studies: The performance of related individuals on tests mainly educative and mainly reproductive.* MSc Thesis, University of London.

Reitman, D., Currier, R. O., & Stickle. T (2002). A critical evaluation of the Parenting Stress Index-Short Form (PSI/SF/SF) in a Head Start population. *Journal of Clinical Child and Adolescent Psychology*, 31, 384-392.

Rodriguez, C. M. & Murphy, L. E. 1997. Parenting stress and abuse potential in parents of children with developmental disabilities. Child *Maltreatment, 2*(3), 245-251.

Roid, G. (2003). *Stanford Binet Intelligence Scales-5th edition.* Chicago: Riverside.

Rosner, B. A. Hodapp, R. M., Fidler, D. J., Sagun, J. N., & Dykens, E. M. (2004). Social competence in persons with Prader-Willi, Williams and Down's syndromes. *Journal of Applied Research in Intellectual Disabilities, 17,* 209-217.

Rosnow, R. L., & Rosenthal, R. (1996). Computing contrasts, effect sizes, and counternulls on other people's published data: general procedures for research consumers. *Psychological Methods, 1,* 331-340.

Saunders, J. A., Morrow-Howell, N., Spitznagel, E., Dore, P, Proctor, E. K. et al. (2006). Imputing missing data: a comparison of methods of social researchers. *Social Work Research, 30*(1), 19-31.

Schoch, K., Powell, K., Callanan, N, Havercamp, S., & Tasse, M. (2006). Behavioral differences in children with Prader-Willi syndrome due to deletion, maternal disomy, and imprinting defects. In Eunpu, D. Brewster, S. (Eds.) *Journal of Genetic Counseling*. *15*(1), 1-26.

Seligman, M. & Darling, R. B. (2007). *Ordinary Families, Special Children* (3rd ed.) New York: The Guilford Press.

Seymour, F. W., Brock, P., During, M. Poole, G. (1989). Reducing Sleep Disruptions in Young Children: Evaluation of Therapist-guided and Written Information Approaches: A Brief Report. *Journal of Child Psychology and Psychiatry 30*(6), 913-918.

Shapiro, N., A., Lessig, M., C., He, A., G., James, G., A., Driscoll, D., J., Lui, Y. (2005) Satiety dysfunction in prader-willi syndrome demonstrated by fMRI *Journal of Neurology, Neuropsychology, and Psychiatry, 76*(2), 260-262.

Simeonsson, R. J., Huntington, G. S., McMillen, J. S., Haugh-Dodds, E., Halperin, D., Zipper, I. N., et al. (1996). Services for young children and families: evaluating intervention cycles. *Infants and Young Children, 9*(2), 31-42.

Spratt, E., G., Saylor, C. F., & Marcias, M., M. (2007) Assessing parent stress in multiple samples of children with special needs. *Families, Systems, and Health, 25*(4), 435-449.

SPSS Inc. (2007). Statistical Package for the Social Sciences, version 17.0. Chicago: SPSS, Inc.

Starr, E. M. (2000). *The Parent Education Perception and Satisfaction Survey: Autism Spectrum Disorders.* Windsor: University of Windsor.

Starr, E. M., Foy, J. B., Cramer, K. M., & Singh, H. (2006). How are schools doing? Parental perceptions of children with Autism Spectrum Disorders, Down Syndrome, and learning disabilities: a comparative analysis. *Education and Training in Developmental Disabilities, 41*(4), 315-332.

Starr, E. M. & Foy, J. B., Cramer, K. M. (2001). Parental perceptions of the education of children with pervasive developmental disorders. *Education and Training in Mental Retardation and Developmental Disabilities, 36*(1), 55-68.

State, M. W., Dykens, E. M. (2000). Genetics of childhood disorders: XV. Prader-Willi syndrome: genes, brain, and behavior. *Journal of American Academy of Child and Adolescent Psychiatry, 39*(6), 797-800.

Suarez, L. M. & Baker, B. L. (1997). Child externalizing behavior and parents' stress: the role of social support. *Family Relations, 46*(4), 373-381.

Tabachnick, B. G., & Fidell, L. S. (2007) *Experimental Design Using ANOVA.* Belmont: Duxbury.

Tabachnick, B. G., & Fidell, L. S. (2007) *Using Multivariate Statistics* (5th ed.). Boston: Pearson Education, Inc.

Tasse, M. J., Havercamp, S. M., & Mandal, R. L. (2002). *Global Assessment of Individual's Behavior–Prader-Willi Syndrome*, version 1.2. University of North Carolina at Chapel Hill.

Tehee, E., Honan, R., & Hevey, D. (2009). Factors contributing to stress in parents of individuals with autistic spectrum disorders. *Journal of Applied Research in Intellectual Disabilities, 22(1),* 34-42.

Thomas, K. C., Morrissey, J. P. & McLaurin, C. (2007). Use of autism services by families and children. *Journal of Autism and Developmental Disorders,* 37, 818-829.

Turnbull, A.P., Turnbull, H. R., Erwin, E. J., & Soodak, L. C. (2006). *Families, Professional, and Exceptionality.* (5th ed.) Upper Saddle River: Pearson/Merrill-Prentice Hall.

U.S. Office of Special Programs. (2000). Study of personal needs in education: the role of paraprofessionals in special education. Ideas that Work; *SPeNSE Fact Sheet.* Retrieved on May 11, 2010 from http://ferdig.coe.ufl.edu/spense/paraFinal.pdf.

University of Michigan Health Systems. (2008). Prader-willi syndrome. T*he University of*

Michigan Medical School. Retrieved on January 1, 2009 from http://www.med.umich.edu/1libr/yourchild/praders.htm#more.

van den Berg, J. E., & Grealish, E. M. (1996). Individualized services and supports through the wraparound process: philosophy and procedures. *Journal of Child and Family Studies, 5*(1), 7-21.

van den Borne, H. W., van Hooren, R. H., van Gestel, M., Rienmeijier, P., Fryns, J. P., et al. (1999). Psychosocial problems, coping strategies, and the need for information of parents with Prader-Willi syndrome and Angelman syndrome. *Patient Education and Counseling, 38,* 205-216.

van den Broek, J. Cunnigham, S. A., Eeckels, R. Herbst, K. (2005) Data cleaning: detecting, diagnosing, and editing data abnormalities. *PLoS Medicine, 10*(2), 966-970.

van Hooren, Widdershoven, van den Borne, & Curfs (2002).Autonomy and intellectual disability: the case of prevention of obesity in Prader-Willi syndrome. *Journal of Intellectual Disability Research*, 46(7), 560-568.

Ware, W. B. (2008). *Statistics/Design III: EDUC 884 Coursepack for Applied Multivariate Analysis.* Chapel Hill, NC: UNC Student Stores Course Pack Publishing.

Westling, D. L. (1997). What parents of young children with mental disabilities want: the views of one community. *Focus on Autism and Other Developmental Disabilities, 12*(2) 67-78.

Wheeler, A. Hatton, D., Reichardt, A., Bailey, D. (2007). Correlates of maternal behaviors in mothers of children with Fragile X syndrome. *Journal of Intellectual Disability Research, 51*(6), 447-462.

White, N. & Hastings, R. P. (2004). Social and professional support for parent of adolescents with severe intellectual disabilities. *Journal of Applied Research in Intellectual Disabilities, 17,* 181-190.

Whitman, B. Y. (2006). Social work interventions: advocacy and support for families. In Butler, M. G., Hanchett, J. M., & Thompson, T. (Eds.), *Management of Prader-Willi Syndrome.* (3rd ed., pp. 426-439). New York: Springer.

Whitman, B. Y. & Jackson, K. (2006). Tools for psychological and behavioral management. In Butler, M. G., Hanchett, J. M., & Thompson, T. (Eds.), *Management of Prader-Willi Syndrome.* (3rd ed., pp. 317-343). New York: Springer.

Whitman, B. Y., & Thompson, T. (2006). Neurodevelopmental and neuropsychological

aspects of Prader-Willi syndrome. In Butler, M. G., Hanchett, J. M., & Thompson, T. (Eds.), *Management of Prader-Willi Syndrome.* (3rd ed., pp. 245-271). New York: Springer.

Whittington, J. Holland, A. (2004). *Prader-Willi syndrome: development and manifestations.* Cambridge: Cambridge University Press.

Whittington, J. Holland, A., Webb, T. (2009). Relationship between the IQ of people with Prader-Willi syndrome and that or their siblings: evidence for imprinted gene effects. *Journal of Intellectual Disability, 53*(5), 411-418.

Whittington, J. Holland, A., Webb, T. Butler, J., Clark, D., & Boer, H. (2004). Cognitive abilities and genotype in a population-based sample of people with Prader-Willi syndrome. *Journal of Intellectual Disability Research, 48*(2), 172-187.

Wigren, M. & Hansen, S. (2005). ADHD symptoms and insistence on sameness in Prader-Willi syndrome. *Journal of Intellectual Disability Research*, 49(6), 449-456.

Wigren, M., Hansen, S. (2003). Prader-Willi syndrome: clinical picture, psychosocial support and current management. *Child: Care, Health, and Development, 29*(6), 449-456.

Wodehouse, G. & McGill, P. (2009). Support for carers of children and young people with developmental disabilities and challenging behavior: what stops it being helpful? *Journal of Intellectual Disability Research, 53*(7), 644-653.

Wyatt, D. A. (2006). A national approach to crisis intervention and advocacy. In Butler, M. G., Hanchett, J. M., & Thompson, T. (Eds.), *Management of Prader-Willi Syndrome.* (3rd ed., pp. 440-447). New York: Springer.

Young, J., Zarcone, J., Holsen, L., Anderson, C., Hall, S. et al. (2006). A measure of food seeking in individuals with Prader-Willi syndrome. *Journal of Intellectual Disability Research, 50*(1), 18-24.

Zarcone, J. Napolitano, D., Peterson, C., Breidbord, J., Ferraioli, S, Caruso-Anderson, M, et al.(2007). The relationship between compulsive behavior and academic achievement across the three genetic subtypes of prader-willi syndrome. *Journal of Intellectual Disability Research, 51*(60), 478-487.